BONE AND JOINT X-RAY DIAGNOSIS

By

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DEDICATED TO

MY WIFE, CHILDREN AND GRANDCHILDREN

Preface

ROENTGEN in 1895 announced the discovery of the miraculous rays which are known universally by his name. The bony skeleton was the first portion of the human body to be examined with the x-rays, one of the earliest roentgenograms being of the hand of his wife. During the early years the principal application of this new adjunct to the field of medicine was in the demonstration of metallic foreign bodies and the diagnosis of fractures, dislocations and other lesions of the bones. Tremendous advances have been made in roentgen diagnosis and therapy and there now remain but few branches of medicine in which x-ray examination has not assumed a position of importance. The skeletal system has continued to be one of the most fertile fields for roentgen study. There are many reasons for this. The bones and joints do not lend themselves to the usual methods of physical examination because of their structure and anatomical positions in the body. Many osseous and articular lesions are difficult or impossible of detection clinically as the manifestations are late in appearing and the signs, symptoms and laboratory data are often obscure and ill-defined, if not actually misleading. Prior to the advent of the x-ray, diagnosis was in many instances a matter of guesswork and became possible only in the advanced stages. Roentgenography, on the other hand, is particularly applicable to the bony structures and is the best method of demonstrating anomalies, injuries, diseases, neoplasms and other lesions of the bones and joints.

In most instances it is possible to identify the disease process, establish its etiology, state whether it is localized or generalized and determine if it is active or quiescent. Roentgen methods of study have prevented much needless suffering, avoided many unnecessary operations and been instrumental in reducing the mortality from many diseases. The x-rays have proven invaluable in the evaluation of surgical and other therapeutic measures, the detection of complications and the prevention of recurrences. They occupy a unique position in the demonstration of the manifestations of numerous disease processes. Certain conditions which were formerly considered rare and of little or no interest since they could be diagnosed only at operation or the autopsy table can now be detected in the incipient stages with the aid of the x-ray, often when the prevention of extension and dissemination is possible. This is of particular significance in neoplasms and infections which are progressive in nature and the only hope of a cure is dependent on early diagnosis.

Roentgenography is of especial aid to those engaged in the diagnosis and treatment of lesions of the bones and joints. It has established orthopedics on a sound basis as a specialty and has resulted in a great

broadening of its scope The orthopedist owes an eternal debt to Roentgen Improvements in equipment refinements in technique and increase in knowledge contribute constantly to the increasing and more widespread utilization of the x ray The importance and value of adequate competent roentgen study cannot be overemphasized The data made available by roentgen study permit, in many cases, of the establishment of the diagnosis earlier and more accurately than by any other clinical method With the exception of histopathologic study it is the most important aid in diagnosis available to the medical profession

The highest standards of excellence must prevail in every phase of the roentgen study Those who undertake the interpretation of roentgenograms bear a great responsibility An incorrect diagnosis may do incalculable harm Failure to detect or properly evaluate the x ray manifestations may result in the conversion of a curable condition into a hopeless disease *Errors of omission are as serious as errors of commission* The value of roentgenography is dependent on the care and completeness of the examination and the diagnostic acumen of the radiologist

The purpose of this book is to describe and discuss the criteria on the basis of which a roentgen diagnosis can be established The potentialities *and scope of the method are evaluated and its limitations discussed* The emphasis is on the description and evaluation of the roentgen manifestations of the diseases and anomalies in which roentgenography is of value In the less common conditions the clinical aspects are discussed for the sake of completeness and to obviate the necessity of referring to other material which may be widely scattered in the literature and not readily available to many readers Radiographs illustrating the principal lesions are reproduced and described

While intended primarily for the roentgenologist and those in training in the field of radiology it will also prove of value to the orthopedist surgeon clinician and others interested in the study of the bones and joints The material is largely from the files of the Boston City Hospital Despite the tremendous advances in the field of roentgenology which have been made in recent years new applications for the use of the x ray are being discovered constantly and diagnostic criteria are being expanded continuously It has been our aim to present the state of current knowledge as completely as possible

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BOSTON MASS

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broadening of its scope. The orthopedist owes an eternal debt to Roentgen. Improvements in equipment, refinements in technique and increase in knowledge contribute constantly to the increasing and more widespread utilization of the x ray. The importance and value of adequate competent roentgen study cannot be overemphasized. The data made available by roentgen study permit, in many cases, of the establishment of the diagnosis earlier and more accurately than by any other clinical method. With the exception of histopathologic study, it is the most important aid in diagnosis available to the medical profession.

The highest standards of excellence must prevail in every phase of the roentgen study. Those who undertake the interpretation of roentgenograms bear a great responsibility. An incorrect diagnosis may do incalculable harm. Failure to detect or properly evaluate the x ray manifestations may result in the conversion of a curable condition into a hopeless disease. Errors of omission are as serious as errors of commission. The value of roentgenography is dependent on the care and completeness of the examination and the diagnostic acumen of the radiologist.

The purpose of this book is to describe and discuss the criteria on the basis of which a roentgen diagnosis can be established. The potentialities and scope of the method are evaluated and its limitations discussed. The emphasis is on the description and evaluation of the roentgen manifestations of the diseases and anomalies in which roentgenography is of value. In the less common conditions the clinical aspects are discussed for the sake of completeness and to obviate the necessity of referring to other material which may be widely scattered in the literature and not readily available to many readers. Radiographs illustrating the principal lesions are reproduced and described.

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BOSTON, MASS.

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HEREDITARY, CONGENITAL AND DEVELOPMENTAL DISTURBANCES OF THE SKELETON

I have utilized numerous articles published in the medical literature by myself often in collaboration with my fellow workers and thank the editors of the Journals in which these papers appeared and my collaborators for the use of the following

Pellegrini Stieda s Disease Amer J Roent *32*, 189-195 1935 With Joseph Resnik

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Bone and Joint X-ray Diagnosis

Chapter

1

General Considerations

INTRODUCTION

THE bony skeleton is one of the large anatomic systems which lends itself readily to study by roentgen methods Roentgen study demonstrates anomalies disease, injuries tumors, and other conditions which affect the bones It is possible to demonstrate lesions in the bones or joints and in most instances to identify them accurately and establish their etiology A large proportion of the problems in the field of x ray diagnosis with reference to the skeleton is concerned with trauma and the demonstration of fractures or dislocations is accurate in practically 100 per cent of cases It is important to stress the value of negative findings in patients who have symptoms referable to the osseous system as the absence of roentgen manifestations rules out practically all of the most serious conditions

The technical aspects of the roentgen examination are of the utmost importance Every roentgenogram must be of the highest standard as regards clarity of outline sharpness of definition, and accuracy of detail In the study of the bones the examination must in every instance comprise at least two views at right angles to each other These usually consist of an anteroposterior and a lateral projection In many instances it is necessary to supplement these by oblique or semilateral, posteroanterior, and other views While this can easily be accomplished in the extremities it is not always possible to obtain lateral projections in certain regions of the body particularly the hip joint the shoulder girdle, the clavicle, and the pelvis In these instances special procedures must be resorted to If the roentgenogram is not absolutely perfect from a technical point of view the observer must not hesitate to ask that the examination be repeated Similarly, if further data can be obtained by additional projections studies with either lighter or darker roentgenograms stereoscopy the use of the Bucky diaphragm or other special procedures such as laminography, these must be made before a final conclusion is reached It must also be stressed that in many instances the first examination may not establish an absolutely definite diagnosis This is particularly true in osteomyelitis and early bone neoplasms Re examinations at a later date may prove invaluable and must be resorted to in all doubtful cases The delayed appearance of fractures is not an uncommon occurrence, particularly

tuberculosis of the hip or after a trauma with prolonged immobilization the term atrophy is also appropriate Hypertrophy means increase in size If one limb is injured or removed, there may be compensatory hypertrophy of its fellow

Correct and careful terminology should be utilized in the requests for roentgen examination as well as in the reports comprising the findings and interpretation of the radiologist Many examples can be offered to illustrate the importance of care in the wording of the x ray requisition In the examination of the wrist after an injury to the carpal bones studies in oblique positions and in ulnar or radial deviation may reveal a fracture which cannot be shown on routine anteroposterior and lateral roentgenograms After an injury to the tarsus the conventional studies of the ankle may prove actually misleading In the case of the knee, the radiologist and technician should know whether a fracture of the patella is suspected as special projections are required for the study of this bone In the search for osteochondritis dissecans of the bones of the knee, intercondylar views are often the only means of demonstrating the lesion The ribs are not always well visualized on the roentgenogram of the chest, the demonstration of fractures being successful only if the affected region is in close apposition to the roentgen film and the direction of the incident beam is correct Studies of the base of the skull the petrous regions, and the mastoid area require special positions which are different from those utilized routinely and it is essential that the clinician indicate the probable site of the lesion in order that the correct roentgen procedures may be used Similar examples may be multiplied endlessly The importance of accuracy and care in indicating the regions which should be studied can not be overemphasized A request for an x ray of the arm is incorrect in a case of Colle s fracture and the same is true if ' the leg ' is asked for when study of the ankle is desired Roentgen examination of the "back ' should not be requested rather the lumbosacral dorsal or cervical spine The expression "take a picture," is inadvertently used on occasion It is much more satisfactory to ask "have roentgen study made or "take x rays of " A request for conventional or routine views is in good order and may be supplemented by special projections if necessary in the opinion of the clinician or the radiologist

It is the proper function of the radiologist to supervise the requisitions for roentgen procedures In large institutions with a frequent turnover of the resident physicians and interns, a member of the staff of the department of radiology can greatly increase the efficiency of the department as well as enhance immeasurably the value and usefulness of the roentgen studies by a careful review of all requisitions for roentgen examination Consultation with the referring physician is advisable in all doubtful matters This is of particular benefit in cases in which the roentgenograms are made by technicians In private practice or after roentgenoscopy, the radiologist instructs the technician as to the procedures and projections which are to be utilized While most clinicians welcome the suggestions of the radiologist and are grateful for his cooperation, there are occasional instances in which the referring physician indicates that he wishes his order carried out exactly as given The radiologist who wishes to serve in the capacity of a true consultant and perform his duties for the optimum benefit of the patient will resist all attempts to relegate him to the position of a technician He quite properly will refuse to do an incomplete, unsatisfactory or otherwise unacceptable examination as being inconsistent

in the flat bones such as the carpal navicular, the ribs, and the spine Post traumatic calcific depositions in the collateral tibial ligaments may not be demonstrable by x ray until two three, or more weeks after an injury In this type of case a single negative exami nation immediately or soon after a trauma cannot be accepted as final proof that no lesion is present, repeated studies on one or more occasions being required to establish the diagnosis

In the roentgen examination of the bones and joints, the roentgenologist is aided greatly if the clinician localizes accurately the area which is to be examined It is essential that the roentgen tube be centered as nearly as possible over the lesion in order to avoid distortion or obscuration of im portant details It will prove definitely more satisfactory if the study is made of a specific area rather than in the form of a general survey Altera tions incidental to the age of the patient are significant and the findings may be dependent in large part on these changes Abnormalities of articular surfaces narrowings of joint spaces and the presence of osteo arthritis may alter the roentgen findings In older people there is frequently osteoporosis or other manifestations of senility Calcification in and about the joints in the walls of the arteries and in other portions of the body may cause confusing shadows or result in the obscuration of important details Significant changes incidental to posture occupation or development are frequent and must be taken into consideration Indi viduals who do heavy work with their arms, legs or trunk may manifest definite alteration in the bones Rickets and other diseases of infancy and childhood may produce curvatures of the spine abnormalities in the vertebræ bowing of the legs and flaring of the ribs which may cause con fusion in diagnosis unless the underlying causes of the changes are appre ciated In the study of the roentgenogram and the demonstration of an abnormality it is helpful to understand whether the condition is general or localized Gross hypertrophic changes may be present in various por tions of the body and Paget s disease may involve multiple bones It is important to establish whether the pathologic process is of a proliferative or a destructive nature New bone formation or increased bone density indicates proliferation or construction of bone This is seen particularly in hypertrophic arthritis, benign and malignant tumors and varying types of chronic bone infection Destructive bone lesions are characterized by loss of tissue or atrophy of the bone This is particularly common in tuberculosis osteomyelitis and malignant tumor

Roentgen Terminology It is important to define the roentgen terms so that it will be clear what the roentgenologist has in mind The terms ' increased radiance or "diminished density mean a change in the bone which permits the roentgen ray to penetrate more easily In an area of increased radiance there is absence or diminution of the normal trabeculæ Because of the lessened amount of lime salts in this area the roentgen ray penetrates more easily and causes a denser shadow on the roentgenogram The terms increased density and ' decreased radiance denote that a change has taken place which has made the affected area more absorptive to the x ray Decreased radiance as applied to the osseous structures indicates an increase in the amount of lime salts or other dense material in the bone The terms atrophy and hypertrophy are frequently used and should be clearly understood Atrophy may refer to the quality or the size of the bones Absorption of lime salts in the bone constitutes atrophy If growth is interfered with or the bone shows lessening of its size as in

in the bone The pathologic and clinical manifestations of the great variety of conditions which are referred for roentgen study must be clearly understood The rare and unusual conditions must be kept constantly in mind as the diagnosis of an obscure condition cannot be established unless the roentgenologist knows of the existence of the disease and the manner in which it manifests itself

THE X-RAY APPEARANCES OF NORMAL BONE

Since bone is more dense than the surrounding soft tissues, its outline and structure are clearly demonstrated by x ray methods of study The calcium content of the bone is the significant factor in the determination of the degree of opacity The outer part of the long bones consists of the cortex which casts a dense roentgen shadow The cortex is wider at the mid-portion of the shaft and becomes narrow toward the end of the shaft The ends of the long bones are composed of a lattice-work of cancellous bone This is less dense than the cortex and presents a characteristic pattern of trabeculation on the roentgenogram Bone marrow occupies the cavities in the bodies of the long bones and the space of the cancellous tissue It is composed principally of fat, areolar tissue, blood vessels, lymphatics, nerves connective tissue and fluid The composition of the marrow is different in the long and flat bones The red marrow occurs in the flat bones and has a high proportion of hematopoietic tissue In old age with disuse, and in certain diseases a large portion of the marrow is replaced by fat The process may be reversed in cases of leukemia, certain of the anemias infections and hemorrhage, the fat in the marrow being replaced by fibrous and other tissue The marrow is less dense than cortical or cancellous bone

The periosteum is a layer of connective tissue and elastic fibers which adheres to the surface of practically every portion of each of the bones except the articular surfaces It has a very important function as it is one of the bone producing elements and its rich supply of finely divided blood vessels penetrates the cortex and nourishes the bone In the normal state the periosteum casts no shadow on the roentgenogram If torn or elevated from the bone hemorrhage takes place and the periosteum becomes clearly visible Periosteum seen on the roentgenogram indicates an abnormality The cortex of the bone is pierced by numerous small channels termed Haversian canals which extend parallel to the axis of the long bone and are united by numerous intersecting canals The Haversian canals serve as paths for the transportation of infection On the roentgenogram the medullary canal appears as a radiant channel as the elements contained within it are less dense than the bone The canal does not traverse the entire length of the bone but merges at its end into the cancellous portion The nutrient foramen is at approximately the middle point of the shaft and is an aperture which allows the large vessels to enter the medullary canal In some bones there are two foramens While the foramens cannot be demonstrated by x ray in most instances they are important as they indicate the site by which metastatic infections and malignancies may gain admittance to the medullary canal The joint cartilage covers the ends of the bone and is not visible on the roentgenogram Similarly, the synovial membrane is not demonstrable No periosteum is present and there are no normal elements which produce bone in the joint Therefore, when bone is present in the joint it is the result either of destruction of cartilage and

with his professional position and not in accordance with the principles of Hippocrates.

THE FIVE PRINCIPAL DENSITIES IN ROENTGENOGRAPHY

Roentgenographically, there are five principal densities depending on the atomic numbers of the structures in the regions under observation. 1. Gaseous substances offer very little obstruction to the passage of x-ray and hence produce very markedly radiant shadows on the roentgenogram. 2. Fatty tissues cause only slight absorption of the rays and are slightly less radiant than the air filled lung. 3. The soft tissue structures and the fluids of the body do not permit x-radiation to pass through to as great an extent as gaseous or fatty substances and hence are more dense on the roentgenogram. The body fluids, blood, lymph and chyle, have densities similar to those of the fleshy tissues. 4. The fourth density is produced by the bony structures and calcific formations which are much less easily penetrated by the roentgen rays due to their high content of calcium and other inorganic substances. 5. The fifth density is produced by radiopaque substances such as barium, iodine containing compounds and similar substances introduced into the body for diagnostic or therapeutic purposes and metallic objects such as missiles and needle fragments. These are more dense than the bony structures.

METHOD OF STUDYING THE ROENTGENOGRAM

It is essential that the radiologist develop and pursue in every instance a careful and complete method of studying the roentgenogram and recording his findings. The observations should not be made haphazardly. A systematic procedure must be established and utilized in every instance in order to obtain completeness and accuracy. Only in this way is it possible to avoid errors of commission or omission. The entire roentgenogram is first reviewed to note the age, sex and any gross abnormalities. Each bone is then observed individually. It is our practice to examine every bone twice. First the eyes scan the outline of the bone. This is followed by complete and thorough observation of the osseous structure and detail. The joints are observed as to width and contour. The soft tissues must also be studied with reference to density, outline and structure. The anatomical location of the changes visualized on the roentgenogram is of the greatest significance. It is essential to note whether the process involves the joint and the adjacent tissues or only the joint. Similarly it is necessary to determine whether the shaft, the metaphysis or the epiphysis alone or in various combinations are affected. If a systematic method of thoroughly studying every portion of the roentgenogram is adhered to in every instance, errors will be eliminated or reduced to a minimum. When a deviation from the normal has been noted, the roentgenologist must determine as accurately as possible whether it is developmental or the result of disease. During childhood the bone is quite different than in the adult and the manifestations of both disease and developmental anomalies may vary greatly in the two groups. A thorough knowledge of the anatomy and the development of the bones, the epiphyses and the epiphyseal centers is a basic requirement. The roentgenologist must know the constituent parts of each bone and joint and the blood and lymph supply in order to comprehend how disease processes reach the bone and progress

are in most instances consequent upon trauma, being best exemplified by fractures which have united with displacement of the fragments The bone undergoes definite changes in order to adapt itself to the deformity There is union and reconstruction of the fragments with the formation of new bone in the callus The compact bone of the fragments undergoes dissolution and is replaced by spongy bone The lines of stress in many bones are curved for adaptation to the function of the bone Immobilization of a joint as in ankylosis leads to adaptive reconstruction, the changes being dependent on the position in which the ankylosis takes place The reconstruction may require many years for completion

Increase in Function Wolff's Law Increase in normal pressure and traction such as occurs in strenuous physical labor or exercise results in localized or generalized strengthening of the skeleton The nature of the process is not clearly understood The bones become heavier and the processes, crests and ridges which serve as attachments of muscles become enlarged, strengthened and roughened It must not be assumed that traction alone serves as a specific stimulus to the deposition of bone as the bones or parts of bone subjected to pressure also become reinforced by the production of new bone This is less obvious and frequently is not recognized Increase of physiologic stress within the limits of tolerance acts as a stimulus to the formation of the bone Functional stresses affect the shapes of the bones and changes in the strength and direction of forces applied to the bones lead to alterations in osseous form and structure Wolff first enunciated this principle and his name has been applied to these phenomena The plates and trabeculæ of each bone are arranged spatially in conformity with the stresses to which the individual bones are subjected This results in the development of maximal strength with a minimum amount of material

Increase in function may lead to osteosclerosis, the formation of more than the normal amount of bone A striking example of this manifestation may occur in kyphosis One portion of a vertebra may be under increased pressure while an adjacent segment may bear little or no weight The part of the vertebra which is under increased pressure presents an increase in the number of spongy trabeculæ, the individual trabeculations are increased in thickness, and the marrow spaces are narrowed The relatively inactive portions of the vertebræ show thinning and diminution of the number of trabeculæ with widening of the marrow spaces Similar changes occur after the healing of a fracture which has not been properly reduced The resultant malunion subjects the fragments to stresses which are abnormal in direction and strength Resorption and rebuilding of the bone ensue with resultant change in the shape of the bone, the arrangement of the freshly formed trabeculæ coinciding with the newly established lines of pressure and tension Similar alterations occur after ankylosis in which the affected bones are in abnormal position It is not clearly understood why the application of mechanical force may stimulate the apposition of bone while lack of function results in bone resorption Bone differs in both macroscopic and microscopic detail according to whether it is normally subjected to tension or pressure Forces within the limits of tolerance of the bone stimulate bone apposition when applied to a particular portion of the bone and in such a direction that they act as intensified normal forces Excessive pressure applied to a segment of bone or forces acting in a direction which deviates from the normal result in bone resorption Roux believed that increased pressure leads to bone apposition only in the regions in which the

exposure of the underlying bone which subsequently proliferates or calcification of the separated synovia

THE REACTION OF BONE TO DISEASE PROCESSES

The reactions of bone which are demonstrable by roentgen study are two in number, bone destruction and bone production The evaluation of the extent of these processes, their location and the progression on serial studies is the basis on which a diagnosis can be established Bone destruction may occur as a result of infection malignancy traumatism, metabolic disturbances and other causes The degree of the reaction depends upon the site of involvement, the virulence of the exciting agent the age of the patient the resistance of the host, and many other factors The point of contact between the irritant or exciting agent and the bone is a site of stimulation It requires considerable time for nature to lay down new bone If the exciting agent is virulent and active the point of contact is swept away and others are formed before there has been time for new bone formation In this case the resultant process is entirely destructive in character If the point of contact remains stationary for a sufficiently long time and the resistance of the host is adequate new bone is formed In many processes such as metastatic carcinoma and myeloma there is continuing destruction without bone production In other instances these processes go on together In osteomyelitis the disease spreads by means of the Haversian canals and their intersecting ramifications Therefore, a characteristic of the disease is its appearance at points distant from the original focus with normal bone areas intervening Primary neoplasms advance by direct extension and new foci are not found separated from the parent growth with normal bone intervening In certain types of neoplasms, new bone occurs within the lesion in areas where normally no bone tissue is present Malignant tumors of the bone which are primary in nature tend to grow in a spherical manner and destroy the cortex by pressure Metastatic tumors are disseminated widely in the bone and there may be normal areas of bone intervening between the areas of bone destruction and proliferation as in osteomyelitis

PRIMARY CHANGES IN THE FORMATION OF BONE

The skeleton may undergo extensive localized or generalized changes in rickets osteomalacia osteitis fibrosa cystica generalisata (Recklinghausen s disease) and many other conditions The resultant deformities may be very important The changes in the long bones under functional stress are adaptive in nature During the healing stages of disease the alterations represent an attempt on the part of the organism to reinforce curved or weakened bones to prevent further bending or fracture In rickets there is progressive deposition of bone at the concavity of the shaft The compact layer becomes thickened and there is partial resorption and replacement by spongy bone A uniformly enlarged marrow space is not present the marrow cavity being partially replaced by bone The compact bone along the convexity usually becomes markedly thinned Concomitantly there is a flattening of the bone in the plane at right angles to the arc of the curve The changes are an adaptation to the altered functional stress associated with the bending of the bone as curved bone has lowered resistance to shearing forces Primary deformities of the skeleton

are in most instances consequent upon trauma, being best exemplified by fractures which have united with displacement of the fragments The bone undergoes definite changes in order to adapt itself to the deformity There is union and reconstruction of the fragments with the formation of new bone in the callus The compact bone of the fragments undergoes dissolution and is replaced by spongy bone The lines of stress in many bones are curved for adaptation to the function of the bone Immobilization of a joint as in ankylosis leads to adaptive reconstruction the changes being dependent on the position in which the ankylosis takes place The reconstruction may require many years for completion

Increase in Function Wolff's Law Increase in normal pressure and traction such as occurs in strenuous physical labor or exercise results in localized or generalized strengthening of the skeleton The nature of the process is not clearly understood The bones become heavier and the processes, crests and ridges which serve as attachments of muscles become enlarged strengthened and roughened It must not be assumed that traction alone serves as a specific stimulus to the deposition of bone as the bones or parts of bone subjected to pressure also become reinforced by the production of new bone This is less obvious and frequently is not recognized Increase of physiologic stress within the limits of tolerance acts as a stimulus to the formation of the bone Functional stresses affect the shapes of the bones and changes in the strength and direction of forces applied to the bones lead to alterations in osseous form and structure Wolff first enunciated this principle and his name has been applied to these phenomena The plates and trabeculæ of each bone are arranged spatially in conformity with the stresses to which the individual bones are subjected This results in the development of maximal strength with a minimum amount of material

Increase in function may lead to osteosclerosis, the formation of more than the normal amount of bone A striking example of this manifestation may occur in kyphosis One portion of a vertebra may be under increased pressure while an adjacent segment may bear little or no weight The part of the vertebra which is under increased pressure presents an increase in the number of spongy trabeculæ the individual trabeculations are increased in thickness and the marrow spaces are narrowed The relatively inactive portions of the vertebræ show thinning and diminution of the number of trabeculæ with widening of the marrow spaces Similar changes occur after the healing of a fracture which has not been properly reduced The resultant malunion subjects the fragments to stresses which are abnormal in direction and strength Resorption and rebuilding of the bone ensue with resultant change in the shape of the bone, the arrangement of the freshly formed trabeculæ coinciding with the newly established lines of pressure and tension Similar alterations occur after ankylosis in which the affected bones are in abnormal position It is not clearly understood why the application of mechanical force may stimulate the apposition of bone while lack of function results in bone resorption Bone differs in both macroscopic and microscopic detail according to whether it is normally subjected to tension or pressure Forces within the limits of tolerance of the bone stimulate bone apposition when applied to a particular portion of the bone and in such a direction that they act as intensified normal forces Excessive pressure applied to a segment of bone or forces acting in a direction which deviates from the normal result in bone resorption Roux believed that increased pressure leads to bone apposition only in the regions in which the

force is transmitted to the bone by intervening cartilage and that pressure directed against endosteum or periosteum results in bone resorption. Pressure exerted physiologically upon bone results in the tissue covering this area differentiating into an avascular tissue. The fact that an area of bone is covered by hyaline cartilage is an indication that it is normally a pressure area.

The law of transformation of bones as expressed by Wolff states that primary changes in form and function are followed by mathematically determinable changes in both the outer shape and the inner architecture of the bone. The law is based on the fact that each bone is fully adapted to the mechanical forces which act upon it. Innumerable observations confirm this law. Deformation of one or more bones may occur in either a normal or a diseased skeleton. In the latter instance the bones may be composed in large part or entirely of uncalcified osteoid tissue as in rickets. This type of bone cannot withstand functional stress and becomes deformed because of its increased physical plasticity. Bones normally show a high degree of rigidity. Changes in functional stress result in deformities because of the inherent biological plasticity of the bone. The growing skeleton has a much greater adaptability to changes in form and function than the adult skeleton. The molding and alteration of the structure of bones are influenced both by growth and normal function. Alteration of function results in a change in the pattern of development of a growing bone and extensive changes in form and structure may take place in a relatively short period of time. In the adult skeleton reconstructive adaptation to abnormal function occurs indirectly and is characterized by structural changes rather than by an alteration in form. The reconstruction of adult bone requires long periods of time, in some instances many years elapsing before the process has been carried to completion.

Lack of Function. Decrease of functional stress results in atrophy or osteoporosis. An example of disuse atrophy is afforded by the changes which take place in bone stumps after amputation. The peripheral segment may atrophy until the shaft actually tapers. The atrophy is accentuated by the lessened or absent pull of the muscles which under normal conditions act on the bone. The changes in an amputation stump are dependent on whether an artificial limb is utilized, particularly in the case of the leg. The tip of the stump may show less reduction in circumference although marked osteoporosis ensues even though an artificial leg is used. The growth of bone is reduced by prolonged disuse. Paralysis of the brachial plexus such as occurs in birth injuries results in shortening of the entire arm. The decrease in growth is in direct ratio to the length of time during which the loss of function persists and it is possible to predict and estimate the reduction of growth of the bones in such cases as the bones grow in an abnormal manner after prolonged disuse.

The Effect of External Forces on the Bone. Cultural customs play an important role. The biologic plasticity of bones is well illustrated in cases in which external forces are employed to mold the growing bone according to an idea fixed by cultural or other tradition. Striking examples are the deformities of the feet of Chinese women and deformations of the skull in certain Indian tribes. In these cases the force is applied by bandaging of a growing individual, the bones being forced to grow into abnormal shapes. The changes in the feet of Chinese females are twofold. The four outer toes are forced under the sole of the foot and the anterior part of the foot assumes a triangular shape. The longitudinal arch of the foot is increased

markedly in curvature and the posterior portion of the calcaneus is brought into alignment with the tibia and fibula The toes are directed sharply downward The malformations are the result of bandaging which is begun at about the age of five years The deformity becomes fully established at the age of fifteen The bones of the foot become deformed as though made of plastic material In consequence of the deformity, there is lessening of functional stress with resultant disuse atrophy The changes are characterized by slenderness and osteoporosis of the bones of the leg and foot Deformations of the head in infancy because of maintenance of a fixed position or after the application of bandages have been found at some time or other in practically every part of the world The custom was first noted in the Americas and the islands of the South Seas In certain parts of the world it still prevails The shape of the head varies from short and broad to long and narrow, the former often being associated with protrusion and the latter with recession of the forehead The appliances utilized apply an external force to the growing skull The brain volume, mental ability and duration of life are not affected despite the fact that the brain may assume an abnormal shape

Habits Deformities of bone may result from habitual activities or positions It is not definitely agreed whether habitual position of an infant can affect the development of the skull in such a manner as to produce a dolichocephalic or brachiocephalic pattern An example which is well substantiated is change in the jaw due to thumb sucking and similar habits In this instance, an abnormal force acts primarily on the teeth, the force being transmitted to the bone The changes are confined principally to the alveolar process, which in the upper jaw protrudes and in the lower jaw retrudes

Therapeutic Appliances Orthopedic appliances are frequently utilized to correct deformities of the skeleton tension and pressure being employed to reshape the bone In consequence of the pressure, there is a biologic reaction of the bone with deformities under the influence of abnormal stresses The resultant change actually represents a manifestation of biologic plasticity and is not merely passively physical in nature

OSTEOLOGY

The skeleton is defined by Webster as the bones collectively of a human being or other vertebrate and comprises the bony or more or less cartilaginous framework which supports the soft tissues and protects the internal organs In the adult human body the skeleton is constituted of a series of bones supplemented in certain regions by pieces of cartilage For purposes of description the skeleton is divided into three separate portions

(1) The axial skeleton which is composed of the following
   a the vertebral column made up of 26 bones
   b the skull made up of 22 bones
   c the hyoid bone
   d the sternum
   e the ribs numbering 24 bones,

(2) The appendicular bones comprising the following
   a the upper extremities made up of 64 bones
   b the lower extremities made up of 62 bones

(3) The auditory ossicles which number 6

Under normal conditions, the adult skeleton numbers 206 bones. The patellas are included in this total, but the smaller sesamoids are not counted. At the time of birth the human body is composed of approximately 270 bones. Fusions of some of these during infancy result in a slight decrease in number. There is a continuing increase in the number of the bones until puberty because of the development of the epiphyses and the bones of the carpus and tarsus. At puberty there are approximately 350 separate bony masses in the human body and this number continues to increase during adolescence. After this period of life, fusions constantly take place and the number is gradually reduced until the final adult quota of 206 is established although in most instances this number is not attained until the middle decades of life.

Classification. The bones are divided into four main categories: (1) long, (2) short, (3) flat, and (4) irregular.

(1) *The Long Bones.* The long bones are those which occur for the most part in the limbs and comprise the humerus, radius, ulna, metacarpals, femur, tibia, fibula, metatarsals and phalanges. Each bone is composed of a body and two extremities. The body, also termed the shaft or diaphysis, is rounded, usually in the form of a cylinder, and contains in its central portion a cavity which is termed the medullary canal. The walls of the bones are made up of dense, compact tissue which tends to be relatively thick in the mid portion of the bone and thinner in the regions of the extremities. Within the medullary canal there are cancellous tissues which are scanty in the middle of the bone and increase in amount towards the ends. In most instances, the long bones are curved, usually in two planes, rather than straight. This results in imparting greater strength to the bones. The extremities of the bones are usually expanded to afford articulations and broad surfaces for muscular attachments. The bones in most instances present at their ends separate centers of ossification which consist of cancellous tissue surrounded by thin compact bone. These are termed epiphyses and are important in the growth and development of the bones.

(2) *The Short Bones.* The carpus and the tarsus are the chief examples of this group, the patella and certain of the sesamoid bones also being included in the category. The short bones combine strength and compactness in association with a limited degree of mobility. They are made up of cancellous tissues covered by a thin outer layer or crust of compact material.

(3) *The Flat Bones.* The scapula and the bones of the skull and face constitute the principal members of this group, others being the sternum, the ribs, the bones of the hip and, according to some authorities, the patellas. The flat bones of the skull and face comprise the occipital, parietal, frontal, nasal, lacrimal and vomer. The cranial bones are characterized by the presence of inner and outer layers of compact tissue termed the tables of the skull. The outer table is thick and tough while the inner, usually called the vitreous table, is thin, dense and brittle. The tissue which intervenes between the tables is cancellous in nature and is called the diploe. In certain portions of the bones of the skull the diploe becomes absorbed with the resultant formation of spaces filled with air between the two tables, the so called accessory sinuses.

(4) *The Irregular Bones.* In this group are included those bones which because of various anatomical peculiarities do not fall into any of the preceding categories. The members of this group comprise the vertebræ

sacrum, coccyx, temporal, sphenoid, ethmoid, zygomatic, maxilla, mandible, palatine, hyoid, and inferior nasal concha. They are made up of cancellous tissue enclosed within a thin crust of compact bone.

The Surfaces of the Bones. The bones present on their surfaces certain prominences or eminences and depressions of two kinds, (1) articular, and (2) non-articular. The principal examples of articular prominences or eminences are those of the heads of the humerus and femur, and of articular depressions are the glenoid cavity of the scapula and the acetabulum of the hip. The non-articular eminences are named according to their form and shape. A broad, rough, even elevation is called a tuberosity, protuberance or process, while a small, rough prominence is termed a tubercle. A sharp, slender, pointed eminence is called a spine, while a narrow, rough elevation which extends for a variable distance along the surface of a bone is designated as a ridge, a crest, or a line. The non-articular depressions vary widely in form and are described as a fossa, pit, depression, groove, furrow, fissure, notch, and other similar terms. The non-articular eminences and depressions increase the extent of the surface which serves for the attachment of ligaments and muscles and are usually well defined in proportion to the muscularity of the individual. The bones present short perforations which are called foramens and longer passages which are termed canals.

Morphogenesis of the Skeleton

The connective tissue, cartilage, and bone are differentiated from the mesenchyme, a diffuse type of mesoderm. With the exception of the flat bones of the face and the cranial vault, the bones of the skeleton in the mammal show first a blastemal, that is a mesenchymal or membranous stage, second, a cartilaginous phase, and lastly the permanent or osseous condition. This is explained by the fact that the bones of the higher vertebrates, which are inherited directly from the cartilaginous skeletons of the lower forms of life, pass through a preliminary cartilaginous stage. However, the bones which have become adapted to the body by the development of newer features such as a large brain, prominent nose, and the palate do not pass through these preliminary developmental stages but develop directly in membrane. The mesenchyme arises primarily from the primitive streak and secondarily from mesodermal segments and the lateral somatic and splanchnic layers. It is a spongy network composed of branching cells which appear to be in contact with each rather than to anastomose. In the areas between the cells are formed open labyrinthine spaces filled with coagulable fluid. The first indications of the formation of the embryo comprise condensations of the mesenchyme into membranous or blastemal rudiments which later differentiate into cartilage and then bone. In very early embryonic life, the mesenchyme acts as an unspecialized packing or filling material between the external and internal layers of epithelium. Subsequently it differentiates into the various tissues and organs. The inert supporting tissues are peculiar in that a ground substance or matrix develops which frequently occupies more space than the cellular elements. The matrix may be fibrous, cartilaginous, or bony. At present, there is no definite agreement as to whether the matrix has its origin within the substances of the cells of the mesenchyme or becomes organized and laid down in the intercellular spaces. In the embryo, bone begins to appear after the seventh week. The bones are divided into two

distinct types (1) the membrane variety and (2) the cartilage bone The mode of histogenesis is identical in each instance A soft, pre osseous tissue composed chiefly of fibers is first differentiated and this subsequently becomes impregnated with a deposition of lime salts

The Development of Membrane Bone The bones of the face and cranium are the principal representatives of this group They develop directly within blastemal or mesenchymal sheets being preceded by primitive connective tissue membrane At one or several central points within this membrane intramembranous ossification begins These centers of ossification are manifested by the appearance of osteoblasts which promptly deposit bone matrix in the form of spicules The spicules fuse in a network or mesh of trabeculæ which continues to spread radially in all directions During the formation of the matrix some of the osteoblasts remain imprisoned as bone cells and are lodged in spaces which are termed lacunæ Subsequent to the formation of the primary centers of ossification the entire primordium becomes enclosed by the periosteum a fibrous membrane which develops as a result of condensation of the mesenchyme The osteoblasts deposit parallel layers or lamellæ of compact bone along the inner aspect of the periosteum This process is termed periosteal ossification and in the case of the skull results in the development of the dense inner and outer tables of the cranium, the mass of spongy bone between the two tables being termed the diploë A great portion of the originally formed bone is provisional in nature and is absorbed, being replaced as the bone continues to grow and assume its final shape During the period of resorption large multinuclear cells the phagocytes or bone destroyers appear These are termed osteoclasts and are giant cells which destroy or dissolve the previously formed bone

The Development of Cartilage Bone Most of the bones of the human body are preceded by a temporary cartilaginous model which is similar in shape to the final, definitive bone The significant characteristic of this method of bone formation is the preliminary destruction of the provisional cartilage as this cartilage must be eliminated before ossification can proceed Because of this the skeletal elements are termed replacement or substitution bones Subsequent to the removal of the cartilage from a given segment of the bone the development is similar to that in membranous bone and ossification occurs both within the eroded cartilage and peripherally beneath the perichondrium The first is termed cartilaginous or endochondral while the second constitutes perichondral or periosteal bone formation It is essential that each of these terms be thoroughly understood The development of endochondral or cartilage type of bone begins with a condensation of the mesenchyme into precartilage Later the cells of this area of condensation become differentiated into chondrocytes which elaborate a hyaline ground substance and form a crude mold of the skeleton The bone trabeculæ first develop in the connective tissue in the region of the mid portion of the bone, the perichondrium and assume a rounded form which encircles the shaft at the site of subsequent degeneration and resorption of the future cartilage The formation of the perichondral tissue and its extension precedes the destruction and elimination of the original cartilaginous model After the central cartilaginous segments resorb an irregular marrow cavity forms and bone deposition commences in the connective tissue of this region and also on the surfaces of the calcified cartilage still remaining In the case of the long bones the development of the shaft continues in two separate areas The peri

chondral collar of bone extends both proximally and distally to the point at which the epiphyseal cartilage will subsequently form There is simultaneously apposition of new bone at the periphery of the collar with resultant increase in the thickness of the bone Apposition of new bone proceeds by periosteal growth and at the same time there is progression of the endochondral bone formation The destruction of cartilage and its replacement by bone lag behind the growth of the cartilaginous elements and at this stage there is no degeneration of the cells or decalcification of the ground substance After the formation of the bone has become well advanced one or more centers of ossification appear at the extremities of the bone in the case of the long or tubular bones In the epiphyseal ends of the cartilaginous bone model, blood vessels are present in the form of branching canals and extend into the cartilage from the perichondrium The earliest manifestation of ossification of the epiphysis comprises degeneration and calcification of the cartilage in the central portion of the future epiphysis The connective tissue in the region of the blood vessels of the cartilage undergoes resorption and is replaced by spongy bone This is of particular significance as the time of ossification is characteristic within certain limits in each bone of the body and is of definite aid in the determination of the skeletal age of an individual

Bone development progresses by the gradual replacement of the cartilaginous portions of the bone There is replacement of the spongy bone with formation of compact bone in the cortex and the development of a uniform marrow space in the central portion of the bone The ends of the shafts of the long bones remain cancellous The extremities are termed the epiphyses the shaft comprises the diaphysis, and the ends of the diaphysis are termed the metaphyses Cartilage which continues to grow without undergoing degeneration and replacement by bone tissue occurs at only two portions of the bone First the extremities remain covered by articular cartilage Second discs of cartilage persist at the ends of the diaphysis and divide the metaphysis from the epiphysis These are termed the epiphyseal cartilages The remaining cartilage undergoes progressive destruction and absorption and is replaced by spongy cancellous bone The periosteal formation of bone occurs simultaneously with the destruction of the cartilage The development of the compact bone in this instance is similar to that which occurs in the formation of the flat bones of the skull and face and is characterized by increased activity of the inner layer of the perichondrium or periosteum There is modeling of the bones into the shape which they will later assume This is accomplished by the formation of grooves and channels and the development of a concentric cylinder with a central axis in the form of a tube containing blood vessels the Haversian system

Growth of Bone A MEMBRANE BONE In the case of the flat or membranous bones, increase in lateral extent takes place by continued ossification from osteoblastic rich connective tissue The process is simpler than in the endochondral bone This is due to the fact that these portions develop by a condensation of the embryonal mesenchyme and are not preformed in cartilage A center of ossification which acts as the nidus develops in the connective tissue Immature, coarse, fibrillar bone forms trabeculae which radiate from the center of ossification to form a circular plate of bone with thin irregular borders Increase in size is by apposition along its inner and outer surfaces and at the borders The marginal ossification in the cranial bones takes place at sites which in the later stages

of development form the sutures In the early phases the bone is entirely spongy in character An outer and inner layer of compact bone develop gradually and the primary coarse fibrillar bone is replaced by mature lamellated bone The rough surfaces become smooth by the apposition of a compact layer Adjacent bones of the skull become demarcated from each by the development of a suture In the early stages the line of separation is wide and is best described as a syndesmosis The connective tissue in the suture is a remnant of the membranous capsule of the brain and serves as a connection between the inner and outer periosteum of the bones, it undergoes gradual reduction in size and eventually may disappear completely The simple straight suture line may become complicated by the formation of interdigitating processes from two or more adjacent bones with the resultant formation of the so called Wormian bones The growth of the membranous bones of the skull may take place on the free surfaces or at the sutures As with the long bones growth in thickness is by the deposition of periosteally formed bone at the surfaces of the bone

B ENDOCHONDRAL BONE THE GROWTH OF THE TUBULAR BONES (1) *Transverse Growth* The shafts of the long bones increase in thickness by the apposition of new layers upon the periosteal surfaces of the shaft The compact layer lining the marrow space undergoes resorption resulting in the maintenance of a relatively constant ratio between the diameter of the shaft and the thickness of the layer of compact bone The circumferential lamellæ formed by periosteal apposition are replaced by Haversian systems within the bone The resorption proceeds to a greater extent than appears necessary to accomplish the elimination of the extra bone in some instances The bone develops by a process of modelling and reconstructive resorption Pressure appears to be an important factor in bringing about the resorption of bone and acts to produce more bone than is necessary to maintain normal pressure relationships with the resultant creation of space and other factors which act as a stimulus to reconstructive repair Hollowing out of the marrow space is associated with alternating periods of resorption and reconstructive apposition along the medullary surfaces of the bone This is termed tubulation

(2) *Longitudinal Growth* During the period of growth the long bones are made up of a shaft or diaphysis and two extremities which are termed epiphyses and serve for articulation with adjoining bones The diaphysis is demarcated from the epiphyses by two plates of hyaline cartilage the epiphyseal plates The articular cartilages cover the free surfaces of the epiphysis Longitudinal growth is achieved by interstitial growth of the epiphyseal and articular cartilages, thickening of the four plates of cartilage resulting in true lengthening of the bone Partial replacement of the growing cartilage plates by bone leads to lengthening of the bony epiphyses rather than to a lengthening of the bone in its entirety During the central resorption and superficial accretion which accompany bone growth both the endochondral osseous tissue and the primitive or early periosteal layer are destroyed In consequence the cancellous bone with its associated red bone marrow is present only at the ends of the long bones In the mid portion or diaphysis of the bone an extensive marrow cavity is present and is occupied by yellow marrow composed chiefly of fat cells In the long bones and the vertebræ the cartilage at each end of the bone grows rapidly and undergoes ossification by progressive extension from the primary center In the interval between birth and puberty, and in some instances even later, osteogenic tissue invades the terminal cartilages and

secondary centers of ossification termed epiphyses form in these regions The cartilaginous plates between the original shaft and the epiphysis develop new bone during the entire period of lengthening of the bone and this in turn is constantly undergoing a modeling process by which the new bone thus laid down is pruned, reformed, and incorporated as an intrinsic part of the cylindrical shaft The growth is principally along the diaphyseal side After the attainment of adult length, the cartilage ceases to proliferate and undergoes ossification The epiphyses then become firmly united to the diaphyses, the line of junction being marked by the epiphyseal line The free end of the epiphysis becomes an articulation and remains cartilaginous throughout life unless affected by trauma, disease, or similar process In the case of the short tubular bones of the hands and feet an epiphysis is present at only one extremity

Modeling of Bone Longitudinal growth is regulated by the metaphyseal cartilage Secondly, the bone is shaped by the periosteum There is a modeling process by which the new bone which has been laid down is incorporated as an intrinsic architectural part of the cylindrical shaft Normally these two factors are synchronized in their action, the new bone created at the metaphyseal cartilage being molded into its final shape by osteoclastic and osteoblastic activity Hunter terms this synchronous action remodeling of bone The newly formed bone shows a waist-like constriction adjacent to the metaphysis and does not form a direct continuation of the old bone Drey is of the opinion that the double contour seen in roentgenograms of the long bones in infants in the first months of life is due to periosteal activity associated with normal modeling He states that the process of modeling which serves to conserve the tubular form at the metaphysis is best observed by superimposing roentgenograms of the limbs of an infant obtained at different ages to show the modeling of the newly created bone within the limits of the old metaphysis in the form of a wasit like constriction Disturbance of the normal process of modeling results in the bone continuing to grow in a straight line In consequence it is broadened in a club like form at the metaphysis and does not acquire the tubular shape which is present in normal bone Failure of modeling is termed under-constriction or under-tubulation It is not characteristic of any single bone disease or process but occurs in multiple diseases of bone in which the normal coordination of cartilaginous growth and periosteal bone shaping become interrupted It is commonly seen in the metaphyseal club like swelling of Gaucher s disease and Cooley's anemia The broadening of the metaphysis is not caused by augmented intramedullary pressure as was previously supposed rather it is due to failure of the normal function of the periosteum

The disturbance of normal calcification of the metaphyseal cartilage in rickets causes a broadening of the metaphysis due to undertubulation This also occurs in osteomyelitis, particularly when resorption of sequestra is beginning At this time the longitudinal growth is preserved and the sequestra are displaced from the metaphysis into the diaphysis The sequestra serve as a source of irritation which disturbs the normal physiology and a change in the process of modeling takes place with resultant undertubulation indicative of the fact that reabsorption of sequestra is taking place After the sequestrum has been absorbed or removed surgically the bone begins to resume its normal shape and undergoes retubulation In certain disease processes, the periosteal cuff does not increase in

of development form the sutures In the early phase the bone is entirely spongy in character An outer and inner layer of compact bone develop gradually and the primary, coarse, fibrillar bone is replaced by mature lamellated bone The rough surfaces become smooth by the apposition of a compact layer Adjacent bones of the skull become demarcated from each by the development of a suture In the early stages the line of separation is wide and is best described as a syndesmosis The connective tissue in the suture is a remnant of the membranous capsule of the brain and serves as a connection between the inner and outer periosteum of the bones, it undergoes gradual reduction in size and eventually may disappear completely The simple straight suture line may become complicated by the formation of interdigitating processes from two or more adjacent bones with the resultant formation of the so called Wormian bones The growth of the membranous bones of the skull may take place on the free surfaces or at the sutures As with the long bones growth in thickness is by the deposition of periosteally formed bone at the surfaces of the bone

B Endochondral Bone The Growth of the Tubular Bones (1) *Transverse Growth* The shafts of the long bones increase in thickness by the apposition of new layers upon the periosteal surfaces of the shaft The compact layer lining the marrow space undergoes resorption resulting in the maintenance of a relatively constant ratio between the diameter of the shaft and the thickness of the layer of compact bone The circumferential lamellæ formed by periosteal apposition are replaced by Haversian systems within the bone The resorption proceeds to a greater extent than appears necessary to accomplish the elimination of the extra bone in some instances The bone develops by a process of modelling and reconstructive resorption Pressure appears to be an important factor in bringing about the resorption of bone and acts to produce more bone than is necessary to maintain normal pressure relationships with the resultant creation of space and other factors which act as a stimulus to reconstructive repair Hollowing out of the marrow space is associated with alternating periods of resorption and reconstructive apposition along the medullary surfaces of the bone This is termed tubulation

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The Joints The joints or articulations occur at the regions where bones meet They are of two types (1) The synarthrosis is the type of joint in which there is restricted mobility (2) The diarthrosis or freely movable form In the former group, the mesenchyme intervening between the bones becomes differentiated into a uniting layer This may take various forms In the case of the cranial bones, the point of juncture is composed of connective tissue and is termed a suture At the pubic symphysis, there is cartilage between the adjacent bones forming a syndesmosis When bony union results, as at the epiphyses, it is called a synostosis The diarthrodial joints are characterized by a prominent joint cavity between two adjacent movable skeletal parts with a ligamentous capsule at the periphery of the joint The joint cavity arises in the fourth month of fetal life, appearing in the clefts of the loose mesenchyme The capsule of the joint is derived from the denser external tissue and is continuous with the periosteum The cells of the inner surface of the capsule become flattened into the form of a false epithelium and form the synovial membrane The ligaments or tendons of the joint become covered by reflections of the synovial membrane and are in reality external to the joint cavity Sesamoid bones such as the patella, develop in relation to both tendons and joints, usually arising in the substance of the primitive joint and in some instances in a cartilaginous plate The bursas are fluid filled sacs at regions of frictional play and are first noted in the later months of fetal life

HISTOLOGY OF BONE TISSUE

Bone tissue is comprised of two permanent elements, the osteocytes which comprise the specialized bone cells and the intercellular substance consisting of fibrils and a calcified cementing substance The cells which participate in bone formation are known as osteoblasts those which cause resorption of bone are termed osteoclasts The osteoblasts and osteoclasts are present only during active stages of bone formation and destruction and constitute transient elements of bone tissue in contradistinction to the osteocytes or permanent elements of bone The osteocyte of fully developed bone is uniform and characteristic in appearance The body of the cell is ovoid and from it extend multiple branching processes or fibrils which interlock with similar extensions from adjacent cell-bodies The osteocytes in the lacunæ and their processes in the canaliculi are contained within the intercellular tissue the ground substance or matrix Collagen fibrils are present in bundles which vary in thickness from 3 to 5 microns The mineral is composed of salts principally calcium phosphate, and is contained within the cementing substance A thin layer of the inter cellular substance lining the bony lacunæ and canaliculi surrounds the osteocytes and their processes and forms the capsule of the osteocytes Osteoblasts are responsible for the formation and calcification of bone matrix In the process of formation of new bone, the osteoblasts form a continuous layer on the surface of the old bone Each osteoblast measures approximately 15 to 20 microns in diameter The nucleus is situated eccentrically and occupies a position in close relation to the aspect of the cell not in contact with the bone In the process of bone formation, certain of the osteoblasts become encircled by the intercellular substance of the bone and become converted into osteocytes

In bone the vital changes are the result of a balance between bone destruction and new bone formation These processes may occur simul-

length at the same rate as the bone in the metaphysis and there is failure of covering at the metaphysis by periosteum. In consequence the bone develops from cartilage and is broadened laterally. This association of longitudinal growth of bone and the length of the periosteum has been described by Keith as the underlying cause of multiple cartilaginous exostoses, the so called diaphyseal aclasis, and is probably the cause of undertubulation. Weiss discusses the broadening of the bone through failure of normal function of the periosteum and shows that this occurs in Paget's disease. He is of the opinion that the theory that the bone is thickened by central swelling and pressure is not correct but that whatever thickening of bone occurs is the result of periosteal deposition. Pathological broadening of bone at the metaphysis is not pathognomonic of any one disease. It is rather the expression of a pathological process leading to a disturbance in the coordination of growth by the metaphysis and the periosteum. This failure in the normal physiological process of modeling may be produced by multiple mechanisms which are of entirely different origin. It is for this reason that in such varied pathological entities as Gaucher's disease, Cooley's anemia, osteomyelitis and rickets similar deformities occur.

The Epiphyses. The epiphyses are of three types: (1) pressure epiphyses which develop at the extremities of the long bones; (2) traction type, affording processes for the insertions of the muscles such as the trochanters of the femurs; and (3) atavistic, which represent a formerly separate bone, as for example, the coracoid process of the scapula. Most bones present more than one center of ossification. In the human body there are a total of more than 800 such centers although more than half of this number do not develop until after birth. Each bone on the average presents four centers of ossification. The centers of ossification appear earlier in females as a rule. The epiphyses in the female appear to unite sooner than in males, and in consequence growth in length ceases earlier by about three years in the female. In the male most of the epiphyses have fused by about the twentieth year of life.

The Nutrient Foramens. The number of ossific centers varies in different bones. In most of the short bones ossification commences at a single point near the center and proceeds toward the surface. In the long bones there is a central point of ossification for the body or diaphysis and one or more for each extremity, the epiphysis. That for the body is the first to appear. The times of union of the epiphyses with the body vary inversely with the dates at which their ossifications began (with the exception of the fibula) and regulate the direction of the nutrient arteries of the bones. Thus the nutrient arteries of the bones of the arm and forearm are directed toward the elbow, since the epiphyses at this joint become united to the bodies before those at the opposite extremities. In the lower limb, on the other hand, the nutrient arteries are directed away from the knee, that is, upward in the femur, downward in the tibia and fibula, and in them it is observed that the upper epiphysis of the femur and the lower epiphyses of the tibia and fibula unite first with the bodies. Where there is only one epiphysis, the nutrient artery is directed toward the other end of the bone, as toward the acromial end of the clavicle, toward the distal ends of the metacarpal bone of the thumb and the metatarsal bone of the great toe, and toward the proximal ends of the other metacarpal and metatarsal bones.

spaces between the trabeculæ communicate with each other throughout the spongy bone. In the compact bone the lamellæ for the most part are arranged in cylindrical systems, each system consisting of a varying number of concentric lamellæ grouped around a narrow axial canal which contains blood vessels and a small amount of loose connective tissue. This system of concentric lamellæ is termed a Haversian system. The Haversian lamellæ are grouped around the Haversian canal. The osteocytes of the Haversian system are arranged along the long axis of the system, the broad surface being parallel to the lamellæ.

The Development and Growth of Bone Tissue. Bone formation is the result of differentiation of connective tissue. The distinction between endochondral and membranous bone formation relates to the development of the bones themselves and not to the development of the bone tissue. It must be stressed that there is only one type of development of bone tissue. The first manifestation of bone formation is the hyalinization or condensation of the semi-fluid cement substance of the connective tissue in the region of the fibrils. The organic substance formed in this manner is called osteoid tissue. The osteoid tissue subsequently undergoes calcification. Calcification does not comprise precipitation of mineral salts in the organic cementing substance of the osteoid tissue. The organic substance appears to undergo a chemical change prior to or during calcification. Osteoid substance is highly resistant to osteoclasis while bone matrix is easily resorbed. The activity of cells of the connective tissue appears to account for the change from osteoid tissue to bone tissue. The exact mechanism of calcification during the change from osteoid tissue to osseous tissue is not clearly understood. The principal component in the calcifying organic substance is calcium phosphate. Immature bone differs markedly from mature bone, the bundles of fibrils in the matrix and the total mineral content of the ground substance being different in character. Immature bone resembles spongy bone in its fundamental characteristics.

The growth of bone consists of the addition of new bone upon the old, each trabeculation of the immature growing by the superimposition of new bone. The new trabeculæ are added to those which have previously developed, mitotic division of osteocytes not being a factor. It is important to stress that there is no interstitial bone growth. The growth of bone tissue occurs by the addition of new bone tissue to existing free bone surfaces, the process being entirely appositional in character. The replacement of immature bone takes place in the following manner. During the growth of the immature coarse fibrillar bone, there occurs simultaneously resorption and replacement by mature lamellated bone. This replacement follows as a distinct phase after osteoblastic resorption. Bone throughout the body develops primarily as spongy bone. When compact bone first appears it is a simple phenomenon. The primary spongy bone consists of evenly distributed trabeculæ. Apposition of concentric lamellæ on the walls of the marrow spaces reduces the space until finally only a small canal remains in the region of the blood vessels.

The Structural Elements of the Skeleton. A bone when considered as an element of the skeleton is comprised of different tissues. In its simplest form, the bone consists of a plate of compact bone enveloped in a differentiated layer of dense connective tissue termed the periosteum. A bone composed of both compact and spongy bone has in addition endosteum and bone marrow, which is the case in the flat bones of the skull. The majority of the bones of the skeleton have a covering of hyaline

taneously or at different times Resorption of bone continues throughout the period of growth of the skeleton and during adult life is found in association with regeneration or reconstruction of bone Bone destruction is an integral part of the biology of bone The removal of bone is accomplished by the organic and inorganic components of the matrix It is believed that the process termed halisterisis or decalcification of living bone necessitates an elevation of local acidity and it has never been shown that such a change in the pH of tissue fluid takes place Resorption of bone begins with the removal of the organic components of the matrix, the proteolytic action of the osteoclasts being the active force which brings it about This results in the liberation of the calcium salts, which are then removed by the body fluids or the macrophages The osteoclasts do not contain calcium salts although calcium salts have been shown to be present in macrophages somewhat removed from the site of the bone resorption The osteocytes frequently undergo degeneration prior to or during the resorption of bone and in some instances become digested by the cells which resorb bone Reversion to fibroblasts or differentiation into osteoblasts or osteoclasts may take place The osteoclasts are giant cells with as a rule many nuclei The average number of nuclei is 12 to 20, although it may vary from 1 to 100 Both osteoblasts and osteoclasts develop by differentiation from the cells of the loose connective tissue, Maximow s undifferentiated mesenchymal cells or the reticular cells The conditions which influence the differentiation of osteoclasts are uncertain Several theories to explain this process have been advanced One possibility is that aging or necrosis of the bone and the osteocytes produces a chemical alteration of the matrix or the tissue fluid in the ground substance of the bone Increase in the pressure in the tissue adjacent to the bone may act as a stimulus for the differentiation of osteoclasts Another theory is that the direct action of certain elements of the blood or tissue fluids is the important factor decrease in the blood calcium level being believed to be a stimulus to bone resorption The parathyroid hormone is considered an active factor in this respect Osteoporosis in disuse atrophy may be explained on the basis of increase of pressure, stasis of the circulation in the marrow and periosteum leading to edema and rise in tissue pressure

Structure of Bone Bone is not a static tissue but rather is in a constant state of flux throughout life internal reconstruction taking place constantly in accordance with the changes in the mechanical requirements and needs of the skeleton These changes are of the utmost importance to safeguard the function and the structure of bone Bone by its very nature must undergo constant perpetual change The osteocytes normally have only a very limited life span and do not undergo regeneration by mitotic division as is the case with other cells of the body As a consequence the portions of the bone which are approaching or have attained the end of their life cycle must be removed and replaced by new bone with young vital osteocytes The process of bone regeneration is the result of the destruction of osteoclasts and the reparative capacity of the osteoblasts

Mature bone is lamellated the bone being laid down in thin layers which measure 4 to 12 microns in thickness The osteocytes lie in the plane of the lamellæ and are situated in and between the lamellæ The lamellæ are arranged differently in spongy or cancellous and compact bone Spongy bone is composed of bars plates or tubules of bone of varying thickness and length joined together to form a three dimensional network The

arteries originate from the periosteal network and enter the bone as arterioles in the multiple Volkmann's canals which open at the outer surface These two systems of the blood supply communicate with each other through numerous anastomoses In the case of endochondral bones which develop from more than one center of ossification, there is an exception to this rule This applies only during the period in which the bony parts are separated by cartilage The veins of the bone follow the arteries The veins begin as extremely wide venous capillaries into which the arteriolar capillaries open abruptly It is important to stress that within the bone the veins do not possess valves There are valves, however, at the points where the larger veins emerge from the bone

The blood supply of the long bones during the period of growth is particularly significant because of the importance of the circulation in the development of inflammatory and necrotic foci and metastatic tumors During the period when the diaphysis is separated from the epiphysis by discs of cartilage, the arteriolar supply of each segment of the bone is independent The shaft is supplied from three sources, the most important of which is the nutrient artery This is a solitary vessel in the long bones, the only exception being the femur, which has two nutrient arteries The nutrient artery perforates the compact layer of the bone obliquely and as it enters the marrow cavity, divides into a descending and an ascending branch After supplying the marrow, the nutrient artery presents terminal branches which enter the metaphyses and in this region anastomoses with the metaphyseal arteries These smaller arteries originate from the arteries of the muscles and ligaments and enter the metaphysis in the region of its circumference The metaphyseal capillary network ends in the region of the epiphyseal cartilage in long, hair-pin shaped loops The arteries of the epiphyses originate from the capsular artery and form a dense network in the epiphysis The course which the capsular arteries follow from their point of contact with the bone until they extend into the epiphysis are dependent on the relation of the line of capsular attachment to the epiphyseal line The arteries supplying the head of the femur are particularly liable to damage as they are exposed along their extended course in the neck of the femur After the disappearance of the epiphyseal cartilage the connections between diaphyseal and epiphyseal arteries are numerous while the anastomosing branches are small As the age of the individual progresses the nutrient arteries become relatively smaller in size This is due to the fact that there is replacement of the hematopoietic red marrow by fatty yellow marrow

AGE DETERMINATION BY STUDY OF THE SKELETON

The human skeleton in its development passes through successive stages of connective tissue, cartilage and bone No portion of the original membranous skeleton persists to adult life Cartilage is present in every portion of the skeleton The duration of the cartilaginous stage is a rough index of the relative speed of development the more rapid the growth the shorter the cartilaginous phase From birth to about the age of four years, the long bones are composed of trabeculæ interspersed with red marrow The marrow cavity becomes fully established at the age of seven years Between twelve and fourteen years, a patch of fat develops in the midportion of the bone and gradually extends toward both extremities The epiphyses undergo similar changes Fatty metamorphosis is

cartilage on some of their surfaces. This serves for articulation with adjacent bones.

The Periosteum. The periosteum is a layer of connective tissue which varies considerably in thickness in the different areas of bone. It is thicker in the regions and surfaces which do not serve as areas of muscular attachment, being particularly thick along surfaces which are covered only by skin and subcutaneous tissue. In these portions of the bone the periosteum is connected only loosely with the bone and can easily be lifted from the bone. An example of this is seen in the case of hemorrhage. The muscles are attached to the bones directly or end on the periosteum. The tendons always have a direct attachment to the bony surface. If the muscles or tendons attach themselves directly to the bone, the connective tissue extends into the bone. These extensions are known as Sharpey's fibers. In these areas the periosteum is lacking and its functions are taken over by the interstitial connective tissue of the muscle or the tendon. Muscles which are attached to the periosteum and therefore only indirectly to the bone are characterized by periosteum which although thin is firmly attached to the bone. The periosteum is composed of two layers. The outer layer is rich in blood vessels and nerves and presents a dense arrangement of collagenous fibers. The inner layer presents loosely arranged fibers with numerous cells and the blood vessels are relatively sparse. During the period when the bones are growing along their periosteal surfaces, there is a continuous layer of osteoblasts lining the bony surface.

The Bone Marrow. In the fetus and infant the large marrow spaces in the shafts of the long bones and the small communicating marrow spaces in the spongy bones are occupied by red or hematopoietic bone marrow. The red marrow in the shafts of the long bones undergoes gradual replacement by yellow or fatty marrow. This replacement begins in the tibia and the fibula, then in the femur, the forearm, the radius, the ulna, and lastly in the humerus. The marrow in both of the epiphyses of the long bones similarly undergoes a gradual transformation into fatty marrow. In the adult hematopoiesis occurs only in the marrow of the flat bones of the skull, the vertebræ, the ribs and the sternum. The red bone marrow is made up of a reticular tissue, the meshes of the tissue being occupied by cells representing all stages of developing erythrocytes and granulocytes. The blood forming elements disappear during the formation of the fatty bone marrow, most of the reticular cells being transformed into adipose cells. The cavities of the marrow are lined with a delicate membrane of connective tissue termed the endosteum.

The Blood Supply of Bones. The blood supply of each bone depends on its arrangement as spongy or compact bone. In the spongy bone, the blood supply is represented by the blood vessels of the marrow spaces. These are approximately equidistant from the surrounding bone trabeculæ. In compact bone the blood vessels occupy the network of the longitudinal Haversian and the connecting Volkmann's canals. The blood vessels in the canals communicate with those in the periosteum and the bone marrow. Most of the blood vessels within the compact bone are capillaries. Arteries and venules are also present and serve to connect the periosteum with the medullary blood vessels of the bone. The blood supply of the bone is derived principally from two sources. Both small and large arteries enter the bone by perforating the compact outer layer and ramify in the bone marrow, being distributed to the bone marrow and the spongy bone. Another group of arteries supplies the compact bone primarily. These

disease result in partial inhibition with derangement of the growing area These examples are cited to illustrate the fact that study of the epiphyses affords valuable data not only as to age and maturation, but also with regard to many disease processes

It is frequently important to establish the age of an individual for medicolegal or other reasons Radiological manifestations cannot be interpreted on the basis of data collected by the anatomists nor can anatomical evidence of union or non-union of epiphyses be correlated with the roentgen findings as the criteria of fusion of the epiphyses differ by the two methods Radiological fusion precedes the disappearance of visible epiphyseal lines in the anatomical specimen The anatomical changes which occur at the end of the growth period sometimes make it difficult for the radiologist to demonstrate the epiphyseal plate of cartilage as a break in the continuity of the bone as the line of the break becomes increasingly thin and its shape more irregular Continuity of the bone pattern at one level may overlap and obliterate a break in continuity at another level The epiphyseal line is often obscured by superimposition of the soft parts The anatomical and roentgen criteria for the determination of age are not similar and must be treated separately

Studies have been made to establish the time of onset of ossification of the epiphyses of the long bones and the primary centers of the small bones in an attempt to construct tables to illustrate the schedule of events in normal growth In the past it has been possible only to make series of observations which contribute to the general picture It is necessary to resort to various computations of averages because of the relative precocity in girls and normal variations in both sexes The production of specific standards for use by the clinician has never proven entirely satisfactory Leonard is of the opinion that a study of the rate of appearance of the ossification centers in the wrist is the best index of maturation and also affords a workable means of determining the presence or absence of metabolic disturbances in childhood He made arbitrary figures to show the earliest and latest age of appearance of each center and utilized these data as a means of detecting the variations which may occur under pathological conditions and to construct an ossification index which is applicable to the various age groups in boys and girls

Vogt and Vickers made a study of osseous growth and development and drew up tables which for individuals up to the age of six and one-half years have proven most satisfactory for general use The studies of Flory and Todd have established useful standards which have won widespread acceptance Stuart made a series of observations at intervals of three months and his diagrams are particularly helpful during the first year of life Sontag, Snell and Anderson suggest that a roentgenogram of one-half of the skeleton preferably the left side of the body be utilized in estimating the skeletal age in persons less than five years old Their tables show the total number of secondary centers of ossification normally visible in the left side of the body at intervals of three months during the first year of life and thereafter at intervals of one year until the age of five years The differences between boys and girls are clearly shown By counting all of the visible secondary centers of ossification and comparing the total with the tables of Sontag and his co workers the skeletal age can be determined It is necessary to include all of the epiphyseal centers in the long bones of the extremities the round bones of the tarsus and carpus the coracoid process and the greater trochanter of the femur To establish the onset

completed at about the age of twenty years the changes paralleling the time of fusion of the epiphyses except in the case of the upper femur The tibia, sternum pelvic bones and vertebrae contain red marrow throughout life The adult distribution is attained at about the age of twenty-five the approximate time of fusion of the epiphyses of these bones The epiphysis is a cartilaginous area which is present at each end of the long bones of the limbs along the superior and inferior aspect of each vertebral body, and in other special sites where processes are required for the attachments of muscles Ossification of the epiphyses takes place from a special center rather than by penetration from the principal center of



Fig 1 New born infant Roentgen study of the new born infant is of value in determining the presence of congenital malformations and other anomalies and is performed routinely in many hospitals

ossification of the bone As a rule the epiphyses which begin to ossify early are late in uniting with the shaft In the utilization of the progress of ossification as indicators of age and bodily maturation certain age limitations are of particular significance From birth to five years the skeletal age is best manifested by the time of appearance of the centers of ossification From five to fourteen years the bony penetration of cartilaginous areas affords an index of the skeletal age In the period between fourteen to twenty-five years the skeletal age is best determined by the union of the epiphyses and diaphyses In certain endocrine disorders bone formation is severely disturbed while in rickets the disturbance is temporary In achondroplasia and dyschondroplasia, slight degrees of

of the cases is there a metatarsal center At two years practically all girls have all the metacarpal centers and 60 per cent have the second metatarsal epiphysis At three years only the center for the fourth metatarsal remains unossified in a significant percentage of cases, and at four and one-half years all of the bones under consideration are ossified in 100 per cent of girls The appearance of these centers in boys is from six to thirteen months later than in girls There is a definite need for separate male and female standards as the rate at which skeletal development proceeds is distinctly more rapid in girls than in boys The relative precocity of skeletal development of girls is already apparent at three months of age and becomes more pronounced as they grow older The skeletal status of thirteen and a half year old girls is not equaled by boys until they are about fifteen and a half years old

Age Determination by Study of the Sesamoid Bones of the Hands The work of Todd also has aided in the elucidation of the process of ossification of the sesamoid bones The pollicis sesamoids begin to ossify at about eleven years and nine months in the case of girls and a year later in boys Girls show a minor progressive acceleration of skeletal maturation

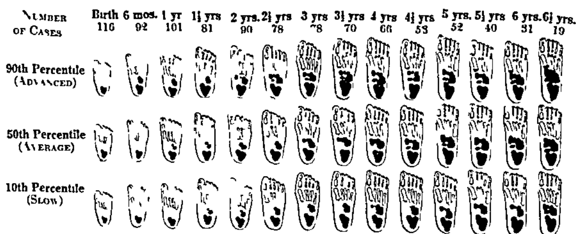


Fig 4 Normal maturation of the bones of the feet in boys (Vogt and Vickers courtesy of Radiology)



Fig 5 Normal maturation of the bones of the feet in girls (Vogt and Vickers courtesy of Radiology)

of adolescence. Buehl and Pyle found the ilium the most satisfactory portion of the skeleton to study as ossification became visible in the iliac crest epiphysis *within 6 months prior* to the onset of the *menarche* in 66⅔ per cent of the girls observed by them. The distal phalanx of the index finger begins to show fusion of the epiphyses with the shaft soon after the onset of the menses. In boys, the iliac center shows visible calcification at 14.5 years, about 1.6 years later than in girls.

Ossification of the Metacarpal and Metatarsal Centers as a Measure of Maturation. Todd pointed out that the primary ossification centers in the carpals, tarsals, metacarpals and metatarsals show more variation than the secondary centers. The only exceptions are the capitate and hamate bones, which ossify within the first three or four months of life. He concluded that the epiphyses of the metacarpals, metatarsals and phalanges are the most useful for determining the bone age, especially in persons under six years of age. Observations were made of 307 boys and *315 girls from birth to five years.* At *one year about 50 per cent of girls* show ossification of the center for the second metacarpal and only 8 per cent have an ossified center for the first metacarpal. In only 4 per cent

Boys

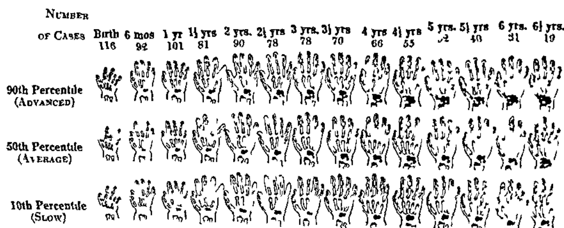


Fig. 2. Normal maturation of the bones of the hands in boys (*Vogt and Vickers, courtesy of Radiology.*)

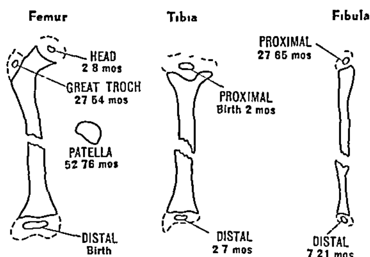
Girls



Fig. 3. Normal maturation of the bones of the hands in girls (*Vogt and Vickers, courtesy of Radiology.*)

girls and between the twelfth and sixteenth year in boys The onset of puberty is predictable with a relatively high degree of accuracy by the time of ossification of the sesamoid at the distal end of the first metacarpal, the sesamoid bone becoming visible in the hand about two years prior to the first menstruation It appears that these bones ossify prior to the beginning of secretory activity of the sex glands and before the onset of the menses The pollicis sesamoids ossify at about the age of ten years in girls and two and one half years later in boys

Serial Studies in Age Determination The time of appearance of the ossification centers in the infant and child is widely used to determine the age and development of the individual and the method has many advan



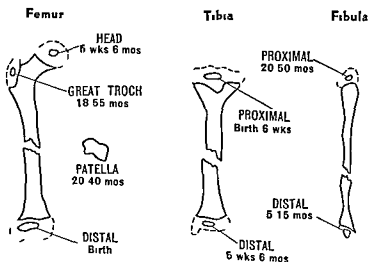
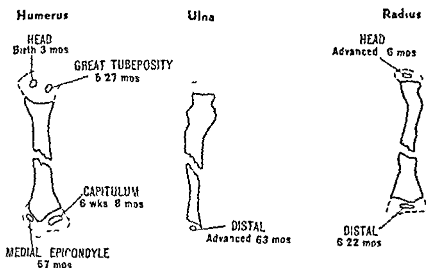


FIG 7 Time of appearance of ossification centers in the femur and lower leg in boys and girls between birth–6½ years (Range indicates 10th and 90th *percentiles*) (Vogt and Vickers courtesy of Radiology)

at the sixth to eighth year of life During the eighth to ninth year there is a deceleration in the maturation of girls and at about the ninth year the degree of maturation is practically equal in the two sexes At the age of ten years there is a period of acceleration in females and by the end of the thirteenth year girls are at the stage attained by boys fifteen years of age After the age of thirteen the maturation of girls again shows a slowing down and at about sixteen and one half years the maturation is approximately equal in both sexes Ossification of the sesamoids may occur from multiple centers and these may later unite or remain permanently separated The sesamoids of the thumb ossify at the age of ten to fourteen in



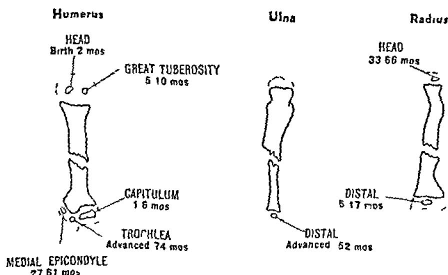


FIG 6 Time of appearance of ossification centers in the arm in boys and girls between birth–6½ years (Range indicates 10th and 90th percentiles) (Vogt and Vickers courtesy of Radiology)

about 1 2 mm during the last six months of the second year As a rule, about 50 per cent of normal children follow the usual pattern, 25 per cent being more rapid and 25 per cent slower than the expected normal The stage of osseous development is well correlated with the height, weight and sexual development Variations are apt to be more pronounced in girls than in boys

Disturbances in osseous development are dependent on many factors and numerous exceptions to the rule may occur Endocrine factors result in more marked alterations than infections Malnutrition must be severe and prolonged to produce demonstrable changes The so called lines of accelerated growth, transverse lines in the metaphyses and diaphyses, may be due to past illnesses, although this is not always the case As a rule, disturbances are most apt to occur during the early years of life and lesions which cause delay in appearance of the centers usually also cause retardation of linear growth of the bones The conditions which are associated with delay in osseous development are hypothyroidism, hypopituitarism, Addison's disease, Frolich's syndrome, chondrodystrophy (achondroplasia), Hurler s syndrome, and mongolism The conditions associated with accelerated osseous development comprise hyperthyroidism, tumors of the adrenal cortex, pituitary basophilism precocious puberty, certain tumors of the ovary such as the granulosa cell, thecoma and teratoma interstitial tumors of the testes, pineal gland tumors in the male, tumors of the third ventricle involving the hypothalamus, polyostotic fibrous dysplasia and simple obesity associated with statural overgrowth

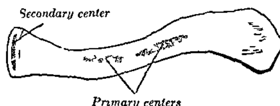


FIG 8 Diagram showing the three centers of ossification of the clavicle (Gray s Anatomy)

THE DEVELOPMENT OF THE BONES

The Hyoid Bone The hyoid is ossified from six centers two for the body, and one for each cornu Ossification commences in the greater cornua toward the end of fetal life, in the body shortly afterward and in the lesser cornua during the first or second year after birth

The Clavicle The clavicle begins to ossify before any other bone in the body, it is ossified from three centers, *viz*, two primary centers, a medial and a lateral for the body which appear during the fifth or sixth week of fetal life and a secondary center for the sternal end, which appears about the eighteenth or twentieth year, and unites with the rest of the bone about the twenty fifth year

The Scapula The scapula is ossified from seven or more centers one for the body two for the coracoid process two for the acromion, one for the vertebral border and one for the inferior angle Ossification of the body begins about the second month of fetal life by the formation of an irregular quadrilateral plate of bone, immediately behind the glenoid cavity This plate extends so as to form the chief part of the bone, the

tages However, the time of appearance of the ossification centers varies considerably and does not necessarily conform to the chronological age in all instances There is much evidence to show that growth and development cannot be judged from the roentgenologic observations of the centers alone Of greater importance than the number and size of the ossification centers at a particular age is the progress of the growth and ossification at intervals of six months or a year This serves as a better index as to whether ossification is normal, retarded or accelerated Also, there must be taken into account the roentgen appearance of the bones in their entirety rather than of the ossification centers alone The comparison of roentgenograms repeated at intervals of six to twelve months is a most important method of judging growth and should be more widely used than it is at present Elgenmath Kelly and Reynolds Christie Maresh, Pyle Currie Levy and others have contributed valuable data to this important field Woolley and McCammon recorded an extensive series of observations of the radius in children during the first two years of life They determined the standard lengths of the radius developed a curve showing the normal rate of growth and emphasized the value of serial roentgenographic observations in the diagnosis and therapy of cases of congenital myxedema Henderson and Sherman observed a series of 100 apparently normal healthy infants and made extensive roentgen studies of the skull in the newborn The importance of these studies cannot be overemphasized as the benefits of observations of this type are very far reaching

The establishment of the stage of osseous development is dependent on the degree of calcification of the component parts of the bones and the growth of the bones Both factors are of importance as they do not always occur simultaneously or at the same rate The appearance and fusion of the various centers of ossification follow a definite and predictable time pattern The observation of this process by roentgen methods permits of estimation of the growth of the individual the skeletal maturity being termed the "bone age In precocious puberty the bone age is greater than the chronological age Prematurity hypothyroidism chronic infections, malnutrition and other conditions may result in retardation of bone development with delay in the ossification of the centers and abnormalities of the sequence of appearance of the centers While the union of the centers may be delayed by various factors the sequence of the union appears to be the same in all individuals There are under normal conditions wide variations in bone maturation racial and sex factors being particularly important There is more rapid growth of the bones in the Negro In infancy the bone age may be either greater or less than the chronological age by several months without necessarily being abnormal The size, contour and density of the component parts of the bones may vary considerably during the period of growth All of the primary ossification centers for the tubular bones become ossified during fetal life The secondary centers of the distal epiphysis of the femur ossify during the eighth to ninth month of fetal life hence absence of this center is important evidence of prematurity The center in the proximal epiphysis of the tibia is ossified in the majority of full term infants at birth The remainder of the secondary centers do not ossify until after birth although one or more carpals normally may be visible at birth The rate of linear growth of the tubular bones affords a more rapid and definite method of appraising the development of the bones than does the maturation of the carpal and tarsal bones The average increase in length is 2 5 mm during the first year and

union it is probable that the detached segment was never united to the rest of the bone

The Sternum The sternum originally consists of two cartilaginous bars, situated one on either side of the median plane and connected with the cartilages of the upper nine ribs of its own side These two bars fuse with each other along the middle line to form the cartilaginous sternum which is ossified from six centers one for the manubrium, four for the body, and one for the xiphoid process The ossific centers appear in the intervals between the articular depressions for the costal cartilages, in the following order in the manubrium and first piece of the body, during the sixth month, in the second and third pieces of the body, during the seventh month of fetal life, in its fourth piece, during the first year after birth, and in the xiphoid process, between the fifth and eighteenth years The centers make their appearance at the upper parts of the segments, and proceed gradually downward Occasionally some of the segments are

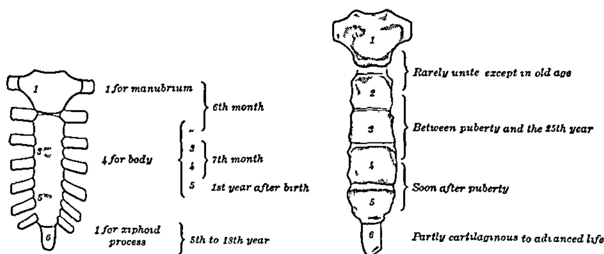


FIG 10 FIG 11

FIG 10 Time of appearance Ossification of the sternum (Gray s Anatomy)

FIG 11 Time of union (Gray s Anatomy)

formed from more than one center, the number and position of which vary Thus the first piece may have three or even six centers When two are present, they are generally situated one above the other the upper being the larger the second piece has seldom more than one, the third, fourth, and fifth pieces are often formed from two centers placed laterally, the irregular union of which explains the rare occurrence of the sternal foramen or of the vertical fissure which occasionally intersects this part of the bone constituting the malformation known as fissura sterni these conditions are further explained by the manner in which the cartilaginous sternum is formed More rarely still the upper end of the sternum may be divided by a fissure Union of the various centers of the body begins about puberty and proceeds from below upward by the age of twenty five they are all united The xiphoid process may become joined to the body before the age of thirty but this occurs more frequently after forty, on the other hand it sometimes remains ununited in old age In advanced life the manubrium is occasionally joined to the body by bone

spine growing up from its dorsal surface about the third month At birth a large part of the scapula is osseous but the glenoid cavity, the coracoid process the acromion the vertebral border and the inferior angle are cartilaginous From the fifteenth to the eighteenth month after birth ossification takes place in the middle of the coracoid process, which as a rule becomes joined with the rest of the bone about the fifteenth year

Between the fourteenth and twentieth years ossification of the remaining parts takes place in quick succession and usually in the following order, first in the root of the coracoid process in the form of a broad scale, secondly, near the base of the acromion thirdly in the inferior angle and contiguous part of the vertebral border fourthly near the extremity of the

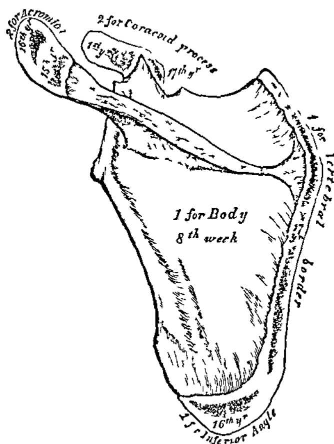


Fig 9 Plan of ossification of the scapula From seven centers (Gray's Anatomy)

acromion fifthly in the vertebral border The base of the acromion is formed by an extension from the spine the two separate nuclei of the acromion unite, and then join with the extension from the spine The upper third of the glenoid cavity is ossified from a separate center (subcoracoid), which makes its appearance between the tenth and eleventh years and joins between the sixteenth and the eighteenth Further, an epiphyseal plate appears for the lower part of the glenoid cavity, while the tip of the coracoid process frequently presents a separate nucleus

These various epiphyses are joined to the bone by the twenty fifth year Failure of bony union between the acromion and spine sometimes occurs the junction being effected by fibrous tissue or by an imperfect articulation in some cases of supposed fracture of the acromion with ligamentous

union it is probable that the detached segment was never united to the rest of the bone

The Sternum The sternum originally consists of two cartilaginous bars, situated one on either side of the median plane and connected with the cartilages of the upper nine ribs of its own side These two bars fuse with each other along the middle line to form the cartilaginous sternum which is ossified from six centers one for the manubrium, four for the body, and one for the xiphoid process The ossific centers appear in the intervals between the articular depressions for the costal cartilages, in the following order in the manubrium and first piece of the body, during the sixth month in the second and third pieces of the body, during the seventh month of fetal life, in its fourth piece, during the first year after birth, and in the xiphoid process, between the fifth and eighteenth years The centers make their appearance at the upper parts of the segments, and proceed gradually downward Occasionally some of the segments are

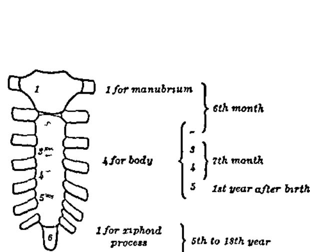


Fig 10

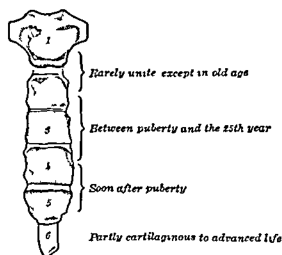


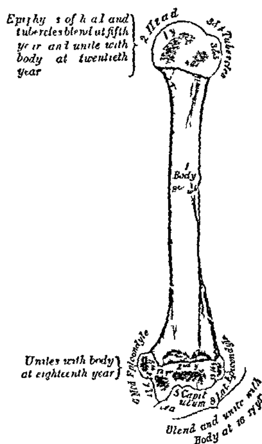
Fig 11

Fig 10 Time of appearance Ossification of the sternum (Gray s Anatomy)

Fig 11 Time of union (Gray s Anatomy)

formed from more than one center the number and position of which vary Thus the first piece may have three or even six centers When two are present they are generally situated one above the other the upper being the larger the second piece has seldom more than one the third, fourth, and fifth pieces are often formed from two centers placed laterally, the irregular union of which explains the rare occurrence of the sternal foramen or of the vertical fissure which occasionally intersects this part of the bone constituting the malformation known as fissura sterni these conditions are further explained by the manner in which the cartilaginous sternum is formed More rarely still the upper end of the sternum may be divided by a fissure Union of the various centers of the body begins about puberty and proceeds from below upward by the age of twenty-five they are all united The xiphoid process may become joined to the body before the age of thirty but this occurs more frequently after forty on the other hand it sometimes remains ununited in old age In advanced life the manubrium is occasionally joined to the body by bone

The Ribs Each rib with the exception of the last two is ossified from four centers, a primary center for the body and three epiphyseal centers, one for the head and one each for the articular and non articular parts of the tubercle The eleventh and twelfth ribs have each only two centers those for the tubercles being wanting Ossification begins near the angle toward the end of the second month of fetal life and is seen first in the sixth and seventh ribs The epiphyses for the head and tubercle make their appearance between the sixteenth and twentieth year and are united to the body about the twenty-fifth year



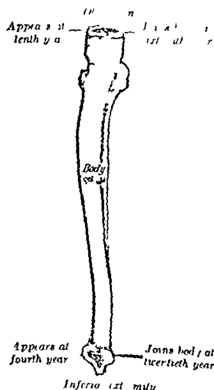


FIG 12 FIG 13

FIG 12 Plan of ossification of the humerus (Gray s Anatomy)

FIG 13 Plan of ossification of the ulna From three centers (Gray s Anatomy)

The Humerus The humerus is ossified from eight centers one for each of the following parts the body the head the greater tubercle the lesser tubercle the capitulum the trochlea and one for each epicondyle The center for the body appears near the middle of the bone in the eighth week of fetal life and soon extends toward the extremities At birth the humerus is ossified in nearly its whole length only the extremities remaining cartilaginous During the first year sometimes before birth ossification commences in the head of the bone and during the third year the center for the greater tubercle and during the fifth that for the lesser tubercle make their appearance By the sixth year the centers for the head and tubercles have joined so as to form a single large epiphysis which fuses with the body about the twentieth year

The lower end of the humerus is ossified as follows At the end of the second year ossification begins in the capitulum, and extends medialward, to form the chief part of the articular end of the bone, the center for the medial part of the trochlea appears about the age of twelve Ossification begins in the medial epicondyle about the fifth year, and in the lateral about the thirteenth or fourteenth year

About the sixteenth or seventeenth year, the lateral epicondyle and both portions of the articulating surface having already joined, unite with the body and at the eighteenth year the medial epicondyle becomes joined to it

The Ulna The ulna is ossified from three centers one each for the body, the inferior extremity, and the top of the olecranon

Ossification begins near the middle of the body, about the eighth week of fetal life, and soon extends through the greater part of the bone At

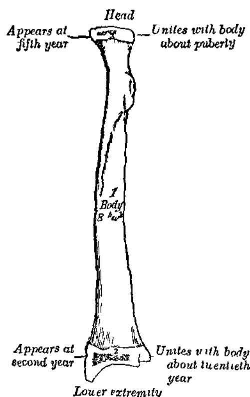
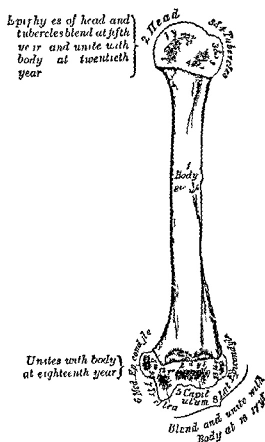


FIG 14 Plan of ossification of the radius From three centers (Gray s Anatomy)

birth the ends are cartilaginous About the fourth year, a center appears in the middle of the head and soon extends into the styloid process About the tenth year, a center appears in the olecranon near its extremity, the chief part of this process being formed by an upward extension of the body The upper epiphysis joins the body about the sixteenth, the lower about the twentieth year

The Radius The radius is ossified from three centers one for the body, and one for either extremity That for the body makes its appearance near the center of the bone during the eighth week of fetal life About the end of the second year, ossification commences in the lower end and at the fifth year in the upper end The upper epiphysis fuses with the body at the age of seventeen or eighteen years the lower about the age of twenty An additional center sometimes found in the radial tuberosity appears about the fourteenth or fifteenth year

The Ribs Each rib with the exception of the last two, is ossified from four centers a primary center for the body and three epiphyseal centers one for the head and one each for the articular and non articular parts of the tubercle The eleventh and twelfth ribs have each only two centers those for the tubercles being wanting Ossification begins near the angle toward the end of the second month of fetal life and is seen first in the sixth and seventh ribs The epiphyses for the head and tubercle make their appearance between the sixteenth and twentieth years and are united to the body about the twenty fifth year



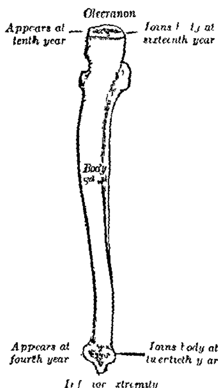


Fig 12 Fig 13

Fig 12 Plan of ossification of the humerus (Gray s Anatomy)

Fig 13 Plan of ossification of the ulna From three centers (Gray s Anatomy)

The Humerus The humerus is ossified from eight centers one for each of the following parts the body the head the greater tubercle the lesser tubercle the capitulum the trochlea and one for each epicondyle The center for the body appears near the middle of the bone in the eighth week of fetal life and soon extends toward the extremities At birth the humerus is ossified in nearly its whole length only the extremities remaining cartilaginous During the first year sometimes before birth ossification commences in the head of the bone and during the third year the center for the greater tubercle and during the fifth that for the lesser tubercle make their appearance By the sixth year the centers for the head and tubercles have joined so as to form a single large epiphysis which fuses with the body about the twentieth year

third metacarpals being the first, and that for the first metacarpal the last to appear, about the third year, the distal extremities of the meta carpals of the fingers and the base of the metacarpal of the thumb begin to ossify, they unite with the bodies about the twentieth year

The phalanges are each ossified from two centers one for the body, and one for the proximal extremity Ossification begins in the body, about the eighth week of fetal life *Ossification of the proximal extremity* commences in the bones of the first row between the third and fourth years, and a year later in those of the second and third rows The two centers become united in each row between the eighteenth and twentieth years In the ungual phalanges the centers for the bodies appear at the distal extremities

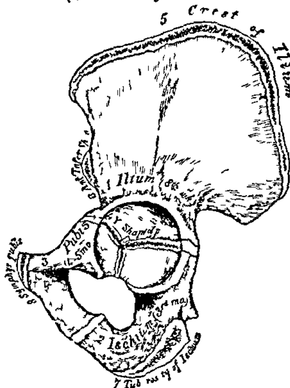


Fig 16 Plan of ossification of the hip bone The three primary centers unite through a Y shaped piece about puberty Epiphyses appear about puberty and unite about the twenty fifth year (Gray s Anatomy)

of the phalanges, instead of at the middle of the bodies, as in the other phalanges Moreover, *of all the bones of the hand, the ungual phalanges are the first to ossify*

The Hip Bone The hip bone is ossified from eight centers three primary —one each for the ilium ischium, and pubis, and five secondary— one each for the crest of the ilium the anterior inferior spine (said to occur more frequently in the male than in the female) the tuberosity of the ischium, the pubic symphysis (more frequent in the female than in the male) and one or more for the Y shaped piece at the bottom of the acetabulum The centers appear in the following order in the lower part of the ilium, immediately above the greater sciatic notch, about the eighth or ninth week of fetal life in the superior ramus of the ischium, about the third month in the superior ramus of the pubis, between the fourth and

The Hand The carpal bones are each ossified from a single center, and ossification proceeds in the following order in the capitate and hamate, during the first year the former preceding the latter in the triangular during the third year in the lunate and greater multangular, during the fifth year, the former preceding the latter in the navicular, during the sixth year, in the lesser multangular, during the eighth year and in the pisiform about the twelfth year Ossification is usually bilaterally symmetrical (Pryor) Occasionally an additional bone the os centrale is found on the back of the carpus lying between the navicular lesser multangular and capitate During the second month of fetal life it is

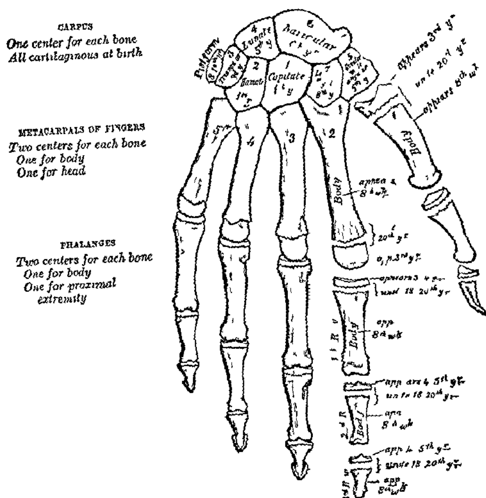


FIG 15 Plan of ossification of the hand (Gray's Anatomy)

represented by a small cartilaginous nodule which usually fuses with the cartilaginous navicular Sometimes the styloid process of the third metacarpal is detached and forms an additional ossicle

The metacarpal bones are each ossified from two centers one for the body and one for the distal extremity of each of the second third fourth and fifth bones one for the body and one for the base of the first metacarpal bone The first metacarpal bone is therefore ossified in the same manner as the phalanges and this has led some anatomists to regard the thumb as being made up of three phalanges and not of a metacarpal bone and two phalanges Ossification commences in the middle of the body about the eighth or ninth week of fetal life the centers for the second and

third metacarpals being the first, and that for the first metacarpal the last to appear, about the third year, the distal extremities of the metacarpals of the fingers and the base of the metacarpal of the thumb begin to ossify, they unite with the bodies about the twentieth year

The phalanges are each ossified from two centers one for the body, and one for the proximal extremity Ossification begins in the body, about the eighth week of fetal life Ossification of the proximal extremity commences in the bones of the first row between the third and fourth years, and a year later in those of the second and third rows The two centers become united in each row between the eighteenth and twentieth years In the ungual phalanges the centers for the bodies appear at the distal extremities

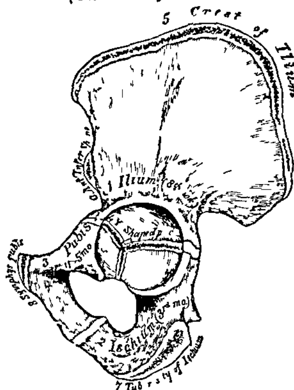


Fig 16 Plan of ossification of the hip bone The three primary centers unite through a Y shaped piece about puberty Epiphyses appear about puberty and unite about the twenty fifth year (Gray s Anatomy)

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The Hip Bone The hip bone is ossified from eight centers three primary—one each for the ilium, ischium, and pubis and five secondary—one each for the crest of the ilium, the anterior inferior spine (said to occur more frequently in the male than in the female), the tuberosity of the ischium the pubic symphysis (more frequent in the female than in the male), and one or more for the Y shaped piece at the bottom of the acetabulum The centers appear in the following order in the lower part of the ilium immediately above the greater sciatic notch about the eighth or ninth week of fetal life in the superior ramus of the ischium, about the third month in the superior ramus of the pubis between the fourth and

fifth months At birth the three primary centers are quite separate, the crest, the bottom of the acetabulum the ischial tuberosity, and the inferior rami of the pubis and ischium being still cartilaginous By the seventh or eighth year the inferior rami of the pubis and ischium are almost completely united by bone

About the thirteenth or fourteenth year the three primary centers have extended their growth into the cartilage which now presents traces of ossification, often by two or more centers One of these the os acetabuli, appears about the age of twelve between the ilium and pubis and fuses with them about the age of eighteen it forms the pubic part of the acetabulum The ilium and ischium then become joined and lastly the pubis and ischium, through the intervention of this Y shaped portion At about

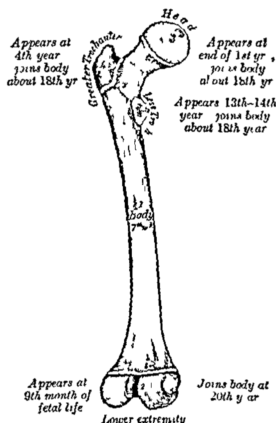


Fig 17 Plan of ossification of the femur From five centers (Gray s Anatomy)

the age of puberty ossification takes place in each of the remaining portions and they join with the rest of the bone between the twentieth and twenty fifth years Separate centers are frequently found for the pubic tubercle and the ischial spine, and for the crest and angle of the pubis

The Femur The femur is ossified from five centers one for the body one for the head one for each trochanter and one for the lower extremity Of all the long bones except the clavicle it is the first to show traces of ossification this commences in the middle of the body at about the seventh week of fetal life, and rapidly extends upward and downward The centers in the epiphyses appear in the following order in the lower end of the bone at the ninth month of fetal life (from this center the condyles and epicondyles are formed) in the head at the end of the first year after birth in the greater trochanter during the fourth year, and in

the lesser trochanter, between the thirteenth and fourteenth years The order in which the epiphyses are joined to the body is the reverse of that of their appearance, they are not united until after puberty, the lesser trochanter being first joined, then the greater, then the head, and lastly, the inferior extremity, which is not united until the twentieth year

The Patella The patella is ossified from a single center, which usually makes it appearance in the second or third year, but may be delayed until the sixth year More rarely, the bone is developed by two centers, placed side by side Ossification is completed about the age of puberty

The Tibia The tibia is ossified from three centers one for the body and one for either extremity Ossification begins in the center of the body,

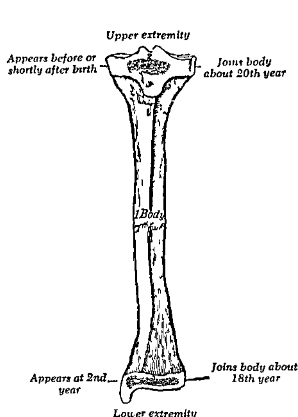


FIG 18

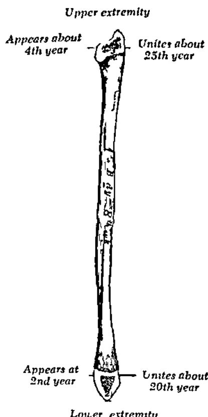


FIG 19

FIG 18 Plan of ossification of the tibia From three centers (Gray s Anatomy)

FIG 19 Plan of ossification of the fibula From three centers (Gray s Anatomy)

about the seventh week of fetal life and gradually extends toward the extremities The center for the upper epiphysis appears before or shortly after birth it is flattened in form and has a thin tongue shaped process in front which forms the tuberosity that for the lower epiphysis appears in the second year The lower epiphysis joins the body at about the eighteenth and the upper one joins about the twentieth year Two additional centers occasionally exist one for the tongue shaped process of the upper epiphysis which forms the tuberosity and one for the medial malleolus

The Fibula The fibula is ossified from three centers one for the body, and one for either end Ossification begins in the body about the eighth week of fetal life and extends toward the extremities At birth the ends are cartilaginous Ossification commences in the lower end in the second

year and in the upper about the fourth year The lower epiphysis the first to ossify unites with the body about the twentieth year the upper epiphysis joins about the twenty fifth year

The Bones of the Foot The tarsal bones are each ossified from a single center excepting the calcaneous which has an epiphysis for its posterior extremity The centers make their appearance in the following order

Calcaneus at the sixth month of fetal life
Talus, about the seventh month

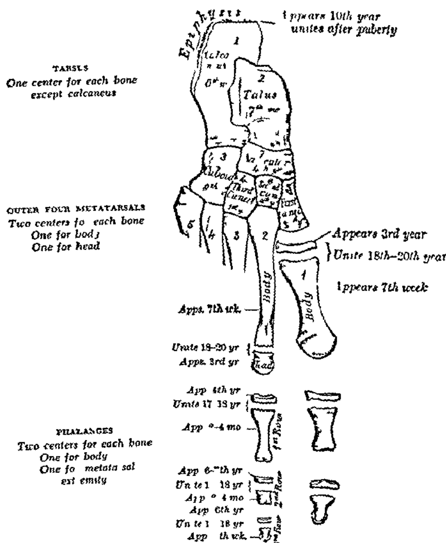


FIG 20 Plan of ossification of the foot (Gray s Anatomy)

Cuboid at the ninth month
Third cuneiform during the first year
First cuneiform in the third year
Second cuneiform and navicular in the fourth year

The epiphysis for the posterior extremity of the calcaneus appears at the tenth year and unites with the rest of the bone soon after puberty The posterior process of the talus is sometimes ossified from a separate center and may remain distinct from the main mass of the bone when it is named the os trigonum The metatarsal bones are each ossified from two centers one for the body and one for the head of the second third

fourth and fifth metatarsals, one for the body, and one for the base of the first metatarsal Ossification commences in the center of the body about the ninth week, and extends toward either extremity The center for the base of the first metatarsal appears about the third year, the centers for the heads of the other bones between the fifth and eighth years, they join the bodies between the eighteenth and twentieth years The phalanges are each ossified from two centers one for the body and one for the base The center for the body appears about the tenth week, that for the base between the fourth and tenth years, it joins the body about the eighteenth year



Fig 21

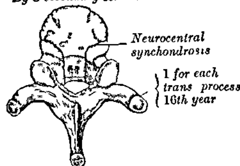


Fig 22

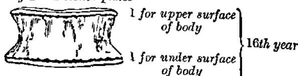


Fig 23

Figs 21–23 Ossification of a vertebra (Gray s Anatomy)

The Vertebral Column Each cartilaginous vertebra is ossified from three primary centers, two for the vertebral arch and one for the body Ossification of the vertebral arches begins in the upper cervical vertebræ about the seventh or eighth week of fetal life and gradually extends down the column The ossific granules first appear in the situations where the transverse processes afterward project, and spread backward to the spinous process forward into the pedicles, and lateralward into the transverse and articular processes Ossification of the bodies begins about the eighth week in the lower thoracic region and subsequently extends upward and downward along the column The center for the body does not

year, and in the upper about the fourth year The lower epiphysis, the first to ossify, unites with the body about the twentieth year the upper epiphysis joins about the twenty fifth year

The Bones of the Foot The tarsal bones are each ossified from a single center, excepting the calcaneus which has an epiphysis for its posterior extremity The centers make their appearance in the following order

Calcaneus at the sixth month of fetal life

Talus, about the seventh month

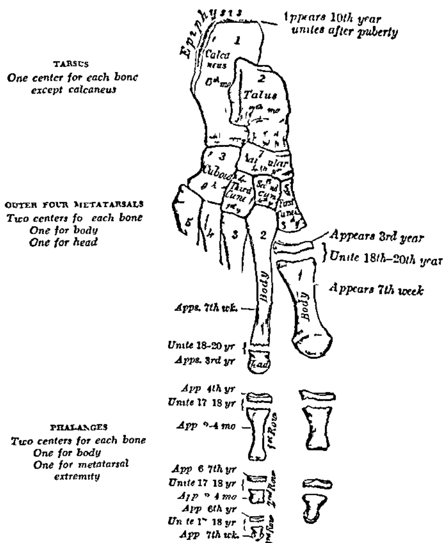


Fig 20 Plan of ossification of the foot (Gray s Anatomy)

Cuboid at the ninth month

Third cuneiform during the first year

First cuneiform in the third year

Second cuneiform and navicular in the fourth year

The epiphysis for the posterior extremity of the calcaneus appears at the tenth year, and unites with the rest of the bone soon after puberty The posterior process of the talus is sometimes ossified from a separate center and may remain distinct from the main mass of the bone when it is named the os trigonum The metatarsal bones are each ossified from two centers one for the body and one for the head of the second third,

fourth and fifth metatarsals, one for the body, and one for the base of the first metatarsal Ossification commences in the center of the body about the ninth week, and extends toward either extremity The center for the base of the first metatarsal appears about the third year, the centers for the heads of the other bones between the fifth and eighth years, they join the bodies between the eighteenth and twentieth years The phalanges are each ossified from two centers one for the body and one for the base The center for the body appears about the tenth week, that for the base between the fourth and tenth years, it joins the body about the eighteenth year



FIG 21

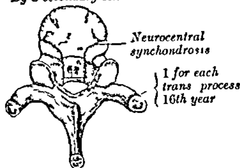


FIG 22

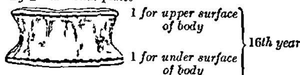


FIG 23

FIGS 21–23 Ossification of a vertebra (Gray s Anatomy)

The Vertebral Column Each cartilaginous vertebra is ossified from three primary centers two for the vertebral arch and one for the body Ossification of the vertebral arches begins in the upper cervical vertebræ about the seventh or eighth week of fetal life and gradually extends down the column The ossific granules first appear in the situations where the transverse processes afterward project and spread backward to the spinous process forward into the pedicles and lateralward into the transverse and articular processes Ossification of the bodies begins about the eighth week in the lower thoracic region and subsequently extends upward and downward along the column The center for the body does not

give rise to the whole of the body of the adult vertebra the postero-lateral portions of which are ossified by extensions from the vertebral arch centers The body of the vertebra during the first few years of life shows, therefore two synchondroses neurocentral synchondroses traversing it along the planes of junction of the three centers In the thoracic region the facets for the head of the ribs lie behind the neurocentral synchondroses and are ossified from the centers of the vertebral arch At birth, the vertebra consists of three pieces the body and the halves of the vertebral arch During the first year the halves of the arch unite behind union taking place first in the lumbar region and then extending upward through the thoracic and cervical regions About the third year the bodies of the upper cervical vertebræ are joined to the arches on either side in the lower lumbar vertebræ the union is not completed until the sixth year Before puberty, no other changes occur excepting a gradual increase of these primary centers the upper and under surfaces of the bodies and the ends of the transverse and spinous processes being cartilaginous About the sixteenth year five secondary centers appear one for the tip of each transverse process, one for the extremity of the spinous process one for the upper and one for the lower surface of the body These fuse with the rest of the bone about the age of twenty five

Exceptions to this mode of development occur in the first, second and seventh cervical vertebræ and in the lumbar vertebræ

ATLAS The atlas is usually ossified from three centers Of these one appears in each lateral mass about the seventh week of fetal life and extends backward at birth, these portions of bone are separated from one another behind by a narrow interval filled with cartilage Between the third and fourth years they unite either directly or through the medium of a separate center developed in the cartilage At birth the anterior arch consists of cartilage in this a separate center appears about the end of the first year after birth and joins the lateral masses from the sixth to the eighth year—the lines of union extending across the anterior portions of the superior articular facets Occasionally there is no separate center the anterior arch being formed by the forward extension and ultimate junction of the two lateral masses sometimes this arch is ossified from two centers one on either side of the middle line

EPISTROPHEUS OR AXIS The axis is ossified from five primary and two secondary centers The body and vertebral arch are ossified in the same manner as the corresponding parts in the other vertebræ *viz*, one center for the body and two for the vertebral arch The centers for the arch appear about the seventh or eighth week of fetal life that for the body about the fourth or fifth month The dens or odontoid process consists originally of a continuation upward of the cartilaginous mass in which the lower part of the body is formed About the sixth month of fetal life two centers make their appearance in the base of this process they are placed laterally and join before birth to form a conical bilobed mass deeply cleft above the interval between the sides of the cleft and the summit of the process is formed by a wedge shaped piece of cartilage The base of the process is separated from the body by a cartilaginous disk which gradually becomes ossified at its circumference but remains cartilaginous in its center until advanced age In this cartilage rudiments of the lower epiphysial lamella of the atlas and the upper epiphysial lamella of the axis may sometimes be found The apex of the odontoid process has a separate center which appears in the second and joins about the twelfth year this

is the upper epiphysial lamella of the atlas. In addition to these there is a secondary center for a thin epiphysial plate on the under surface of the body of the bone.

THE SEVENTH CERVICAL VERTEBRA. The anterior or costal part of the transverse process of this vertebra is sometimes ossified from a separate center which appears about the sixth month of fetal life and joins the body and posterior part of the transverse process between the fifth and sixth years. Occasionally the costal part persists as a separate piece, and, becoming lengthened lateralward and forward, constitutes what is known as a cervical rib. Separate ossific centers have also been found in the costal processes of the fourth, fifth and sixth cervical vertebræ.

LUMBAR VERTEBRÆ. The lumbar vertebræ have each two additional centers, for the mammillary processes. The transverse process of the first lumbar is sometimes developed as a separate piece, which may remain permanently ununited with the rest of the bone, thus forming a lumbar rib—a peculiarity, however rarely met with.

SACRUM. The body of each sacral vertebra is ossified (Figs. 27–30) from a primary center and two epiphysial plates, one for its upper and another for its under surface, while each vertebral arch is ossified from two centers. The anterior portions of the lateral parts have six additional centers, two for each of the three vertebræ; these represent the costal elements and make their appearance above and lateral to the anterior sacral foramina.

On each lateral surface two epiphysial plates are developed, one for the auricular surface, and another for the remaining part of the thin lateral edge of the bone.

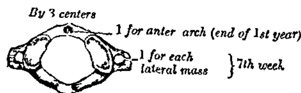


FIG. 24

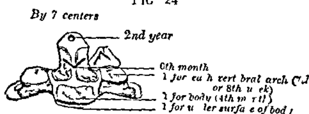


FIG. 25

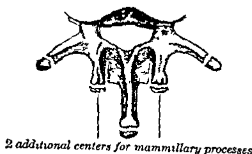


FIG. 26

FIG. 24. Atlas. (Gray's Anatomy.)

FIG. 25. Axis. (Gray's Anatomy.)

FIG. 26. Lumbar vertebra. (Gray's Anatomy.)

PERIODS OF OSSIFICATION About the eighth or ninth week of fetal life, ossification of the central part of the body of the first sacral vertebra commences, and is rapidly followed by deposit of ossific matter in the second and third ossification does not commence in the bodies of the lower two segments until between the fifth and eighth months of fetal life Between the sixth and eighth months ossification of the vertebral arches takes place, and about the same time the costal centers for the lateral parts make their appearance The junction of the vertebral arches with the bodies takes place in the lower vertebræ as early as the second year, but are not affected in the uppermost until the fifth or sixth year About the sixteenth year the epiphyseal plates for the upper and under surfaces of the bodies are formed and between the eighteenth and twentieth years those for the lateral surfaces make their appearance The bodies of the sacral vertebræ are during early life, separated from each other by intervertebral fibrocartilages but about the eighteenth year the two lowest segments become united by bone, and the process of bony union gradually extends upward with the result that between the twenty-fifth and thirtieth years of life all the segments are united On examining a sagittal section of the sacrum the situations of the intervertebral fibrocartilages are indicated by a series of oval cavities

COCCYX The coccyx is ossified from four centers, one for each segment The ossific nuclei make their appearance in the following order in the first segment between the first and fourth years in the second between the fifth and tenth years, in the third between the tenth and fifteenth years in the fourth between the fourteenth and twentieth years As age advances the segments unite with one another, the union between the first and second segments being frequently delayed until after the age of twenty five or thirty At a late period of life, especially in females the coccyx often fuses with the sacrum

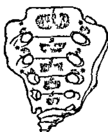


FIG 27

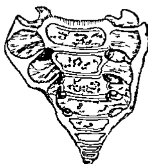


FIG 29

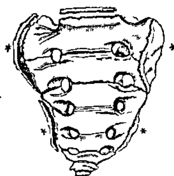


FIG 28

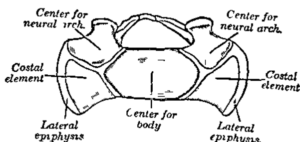


FIG 30

FIGS 27–30 Ossification of the sacrum (Gray s Anatomy)

TIME SCHEDULE FOR THE APPEARANCE AND FUSION OF THE OSSIFICATION CENTERS (GRAY'S ANATOMY)

SKULL

	Appears	Fuses
Occipital Bone		
Squama		
Planum occipitale		
Midline (2)	2nd month fetal	
Away from midline (2)	3rd month fetal	3rd month fetal
Planum nuchale (2)	7th week fetal	
Lateral (2)	8th week fetal	4th year
Basilar	6th week fetal	6th year
Union with Sphenoid		18th–25th year
Parietal Bone	8th week fetal	
Frontal Bone		
Primary centers supraorbital	2nd month fetal	8th year
Temporal Bone		
Squama (including zygomatic process)	2nd month	Before Birth
Petromastoid	5th–6th month	1st year
Prootic		
Opisthotic		
Pterotic		
Epiotic		
Tympanic Ring	3rd month	1st year
Styloid Process		
Tympanohyal	Before birth	
Stylohyal	After birth	After puberty
Sphenoid Bone		
Presphenoid		
Orbitosphenoids (lesser wings)	9th week fetal	
Body	After 9th week fetal	
Sphenoidal conchæ (4)	5th month fetal	9th–12th years
Union with labyrinth of ethmoids		4th year
Postsphenoid		
Alisphenoids (greater wings)	8th week fetal	1st year
Body	After 8th week	Middle fetal life
Medial pterygoid plates	9th–10th week	6th month
Hamulus	3rd month	3rd month
Lingula	4th month	4th month
Ethmoid Bone		
Perpendicular Plate	1st year	2nd year
Labyrinths (2)	4th–5th month fetal	2nd year
Nasal Bones	3rd month fetal	
Lacrimal Bone	12th week	
Maxillæ	6th week fetal	3rd month
Vomer	8th week fetal	3rd month
Inferior Nasal Concha	5th month fetal	
Palatine Bone	6th–8th week fetal	
Zygomatic Bone	8th week fetal	5th month fetal
Mandible	6th week fetal	1st year
Hyoid Bone		
Greater cornua	Near 9th month fetal	
Body	Shortly after birth	
Lesser cornua	1st or 2nd year	

TIME SCHEDULE FOR THE APPEARANCE AND FUSION OF THE OSSIFICATION CENTERS (*Continued*)

	Appears	*Fuses*
Sternum		
Manubrium	6th month fetal	25th year
Body		
First part	6th month fetal	25th year
Second and third parts	7th month fetal	25th year
Fourth part	1st year	25th year
Xiphoid	5th–18th year	Bef[illegible] [illegible] after 40 [illegible]
Ribs	2nd month fetal	
Body		
Tubercule and head	16th—20th year	[illegible]th year
UPPER EXTREMITY		
Clavicle		
Body		
Medial	5th–6th week fetal	[illegible]th year
Lateral	5th–6th week fetal	25th year
Sternal End	18th–20th year	25th year
Scapula		
Body	2nd month fetal	
Coracoid Process (?)	15th–18th month	15th year
Acromion (2)	14th–20th year	25th year
Vertebral border	11th–20th year	25th year
Inferior angle	14th–20th year	25th year
Humerus		
Body	8th week fetal	20th year
Head	1st year	6th year
Greater tubercle	3rd year	6th year
Lesser tubercle	5th year	6th year
Capitulum	2nd year	16th–17th year
Trochlea	12th year	16th–17th year
Medial epicondyle	5th year	18th year
Lateral epicondyle	13th–14th year	16th–17th year
Radius		
Body	8th week fetal	17th–18th year
Proximal	5th year	17th–18th year
Distal	2nd year	20 years
Ulna		
Body	8th week fetal	16th year
Olecranon	10th year	16th year
Styloid	4th year	20th year
Hand		
Capitate	1st year	
Hamate	1st year	
Triangular	3rd year	
Lunate	5th year	
Greater multangular	5th year	
Navicular	6th year	
Lesser multangular	8th year	
Pisiform	12th year	
1st metacarpal		
Body	8th–9th week fetal	
Distal	3rd year	20th year

TIME SCHEDULE FOR THE APPEARANCE AND FUSION OF THE OSSIFICATION CENTERS (*Continued*)

Upper Extremity (*Continued*)

	Appears	*Fuses*
2nd metacarpal		
Body	8th–9th week fetal	
Distal	3rd year	20th year
3rd metacarpal		
Body	8th–9th week fetal	
Distal	3rd year	20th year
4th metacarpal		
Body	8th–9th week fetal	
Distal	3rd year	20th year
5th metacarpal		
Body	8th–9th week fetal	
Distal	3rd year	20th year
Phalanges		
Body	8th week fetal	
Proximal	3rd and 4th year	18th and 20th year
Second	5th year	18th and 20th year
Third	5th year	18th and 20th year
Sesamoids thumb		18th and 20th year

Lower Extremity

Femur		
Head	1st year	Puberty
Body	7th week fetal	Puberty
Greater trochanter	4th year	Puberty
Lesser trochanter	13th–14th year	Puberty
Lower extremity	9th month fetal	20th year
Tibia		
Upper extremity	Birth	20th year
Body	7th week fetal	
Lower extremity	2nd year	18th year
Fibula		
Upper extremity	4th year	25th year
Body	8th week fetal	
Lower extremity	2nd year	20th year
Patella	2nd–3rd year	Puberty
Foot		
Calcaneus	6th month fetal	Puberty
Posterior extremity	10th year	
Talus	7th month fetal	
Cuboid	9th month fetal	
First cuneiform	3rd year	
Second cuneiform	4th year	
Third cuneiform	1st year	
Navicular	4th year	
1st Metatarsal	3rd year	18th–20th year
2nd Metatarsal	5th–8th year	18th–20th year
3rd Metatarsal	5th–8th year	18th–20th year
4th Metatarsal	5th–8th year	18th–20th year
5th Metatarsal	5th–8th year	18th–20th year
Phalanges		
Body	10th week	18th year
Base	4th–10th year	

TIME SCHEDULE FOR THE APPEARANCE AND FUSION OF THE OSSIFICATION CENTERS (*Continued*)

	Appears	*Fuses*
Sternum		
Manubrium	6th month fetal	25th year
Body		
First part	6th month fetal	25th year
Second and third parts	7th month fetal	25th year
Fourth part	1st year	25th year
Xiphoid	5th–18th year	Before [illegible] after 40 [illegible]
Ribs	2nd month fetal	
Body		
Tubercule and head	16th—20th year	25th
UPPER EXTREMITY		
Clavicle		
Body		
Medial	5th–6th week fetal	25th year
Lateral	5th–6th week fetal	25th year
Sternal End	18th–20th year	25th year
Scapula		
Body	2nd month fetal	
Coracoid Process (2)	15th–18th month	15th year
Acromion (2)	14th–20th year	25th year
Vertebral border	14th–20th year	25th year
Inferior angle	14th–20th year	25th year
Humerus		
Body	8th week fetal	20th year
Head	1st year	6th year
Greater tubercle	3rd year	6th year
Lesser tubercle	5th year	6th year
Capitulum	2nd year	16th–17th year
Trochlea	12th year	16th–17th year
Medial epicondyle	5th year	18th year
Lateral epicondyle	13th–14th year	16th–17th year
Radius		
Body	8th week fetal	17th–18th year
Proximal	5th year	17th–18th year
Distal	2nd year	20 years
Ulna		
Body	8th week fetal	16th year
Olecranon	10th year	16th year
Styloid	4th year	20th year
Hand		
Capitate	1st year	
Hamate	1st year	
Triangular	3rd year	
Lunate	5th year	
Greater multangular	5th year	
Navicular	6th year	
Lesser multangular	8th year	
Pisiform	12th year	
1st metacarpal		
Body	8th–9th week fetal	
Distal	3rd year	20th year

CHRONOLOGICAL ORDER OF APPEARANCE AND FUSION OF OSSIFICATION CENTERS (*Continued*)

Before Birth (Continued)

Estimated age 12 weeks	Semicircular canals (Hill)
109 mm (15 weeks)	Bodies lower five cervical and third sacral spines arch first sacral middle row of phalanges of fingers ischium distal phalanges of second third fourth and fifth toes
134 mm (17 weeks)	Male Cervical rib
147 mm (18 weeks)	Arch second sacral Male—body fourth sacral
150 mm (18½ weeks)	Female—arch third and fourth sacral Body fourth sacral
165 mm (20 weeks)	Bodies atlas epistropheus Female—Body fifth sacral cervical rib Male—Arch third and fourth sacral pubis calcaneus talus
Estimated age 20 weeks	Hyoid (Hill)
171 mm (20½ weeks)	Male—arch fifth sacral middle phalanx third toe
180 mm (21 weeks)	First second and third sternebræ Male—first sacral lateral mass
185 mm (21½ weeks)	Male—middle phalanx of second toe
205 mm (23 weeks)	Female—arch fifth sacral pubis calcaneus talus
218 mm (25 weeks)	Fourth sternebra
220 mm (25 weeks)	Female—first second and third sacral lateral mass middle phalanges of second and third toes
233 mm (26½ weeks)	Female—middle phalanx of fourth toe
262 mm (30 weeks)	Distal epiphysis of femur Female—first segment of coccyx
275 mm (31 weeks)	Male—middle phalanx of fourth and fifth toes
283 mm (32 weeks)	Fifth sternebra
295 mm (33 weeks)	Sixth sternebra proximal epiphysis of humerus and tibia Male—first segment of coccyx and cuboid
312 mm (35 weeks)	Male—third sacral lateral mass
315 mm (36 weeks)	Female—cuboid

After Birth

First Year	Capitate hamate epiphyses for head of femur distal tibia and fibula center for third cuneiform
Female	Epiphysis greater tubercle humerus capitulum distal radius proximal phalanges of middle ring and little fingers distal phalanx of thumb
Male	Cuboid
Second Year	Epiphyses second and third metacarpals proximal phalanx of index
Female	Epiphyses first metacarpal proximal phalanx of thumb middle phalanges of middle and ring fingers distal phalanges of middle and ring fingers centers tarsal navicular first and second cuneiforms epiphyses proximal phalanges of second third and fourth toes middle phalanx third toe
Male	Capitulum epiphyses distal radius proximal phalanges of middle ring and little fingers
Age Two	Epiphysis fourth metacarpal
Female	Epiphyses fifth metacarpal middle phalanges of index and little fingers center patella epiphysis first metatarsal proximal phalanges of hallux and fifth toes middle phalanges of second and fourth toes
Male	Epiphyses greater tubercle of humerus first metacarpal distal phalanges of thumb and index center for first cuneiform epiphyses for proximal phalanges of second third and fourth toes

TIME SCHEDULE FOR THE APPEARANCE AND FUSION OF THE OSSIFICATION CENTERS (Continued)

Vertebral Column

	Appears	*Fuses*
Cervical		
Arches	7th–8th week fetal	3rd year
Body		
Atlas		
Lateral masses	7th week fetal	3rd–4th year
Anterior arch	1st year	6th–8th year
Epistropheus (Axis)		
Arch (2)	7th–8th week fetal	
Body	4th–5th month fetal	
Odontoid		
Base	6th month fetal	Before birth
Apex	2nd year	12th year
7th cervical	6th month fetal	5th–6th year
Thoracic		
Lower	After 6th–8th week	5th week
Arches		1st year
Lumbar	6th–8th week	6th year
Transverse Processes all regions	16th year	25th year
Sacrum		
Body	16th–20th year	18th–25th year
Central 1st 2nd and 3rd sacral vertebræ	8th–9th week fetal	5th–6th year
Lower 2 segments	5th–8th month fetal	2nd year
Arches		6th–8th month
Costal centers for lateral parts	6th–8th month	
Coccyx		
First segment	1st–4th year	25th or 30th year
Second segment	5th–10th year	Advancing age
Third segment	10th–15th year	Advancing age
Fourth segment	14th–20th year	Advancing age
Hip Bone		
Lower ilium	8th–9th week fetal	7th–8th year
Superior ramus ischium	3rd month	7th–8th year
Superior ramus pubis	4th–5th months	18th year
Acetabulum	12 years	18 years

Chronological Order of Appearance and Fusion of Ossification Centers

(Flecker Am J Roent)

Before Birth

(Actual observations only)

30 mm (9 weeks)	Clavicle
32 5 mm (49 days after single coitus)	Proximal 10 pairs of ribs primary centers of scapula humerus radius ulna ilium femur tibia and fibula
62 mm (11½ weeks)	Arches cervical spines bodies and arches thoracic and lumbar spines bodies first and second sacral eleventh pair of ribs primary centers metacarpals proximal and distal rows of phalanges of fingers metatarsals proximal row of phalanges of toes

CHRONOLOGICAL ORDER OF APPEARANCE AND FUSION OF OSSIFICATION CENTERS (*Continued*)

After Birth (Continued)

Age Fourteen	
Female	Epiphyses acromion iliac crest lesser trochanter Union epiphyses medial epicondyle olecranon proximal radius proximal phalanges middle ring and little fingers distal phalanges index middle and ring fingers proximal femur greater trochanter distal tibia and fibula apophysis calcaneus epiphyses all metatarsals proximal phalanges all toes
Male	Union distal epiphyses of humerus to each other
Age Fifteen	Union epiphysis distal phalanx fourth toe
Female	Centers for sesamoids of index and little fingers Union of epiphyses first metacarpal proximal phalanges thumb and index distal phalanx little finger proximal tibia middle phalanx second distal phalanx hallux
Male	Epiphysis for acromion Union of ilium and ischium with pubis distal phalanges second and third toes
Age Sixteen	Centers for distal sesamoids of thumb
Female	Epiphysis tuber ischii Fusion four inner metacarpals middle row of phalanges of fingers
Male	Epiphysis iliac crest Fusions distal conjoint epiphysis to shaft of humerus medial epicondyle olecranon proximal radius distal phalanx middle finger greater trochanter
Age Seventeen	Union of epiphysis acromion
Female	Clavicle Union proximal conjoint epiphysis to shaft of humerus distal ulna distal femur proximal fibula
Male	Fusion epiphyses proximal phalanges thumb middle row phalanges of fingers distal phalanges thumb index ring and little fingers proximal femur distal tibia and fibula apophysis of calcaneus epiphyses all metatarsals proximal row phalanges of toes middle phalanx second toe, distal phalanx hallux
Age Eighteen	
Female	Union epiphyses clavicle distal radius
Male	Center for sesamoid of little finger Fusion epiphyses proximal humerus to shaft all metacarpals proximal phalanges index middle ring and little finger proximal tibia
Age Nineteen	
Male	Center for sesamoid of index epiphysis tuber ischii Fusions distal radius and ulna femur proximal fibula
Age Twenty	
Male	Fusion tuber ischii
Age Twenty one	Fusion iliac crest
Female	Fusion tuber ischii
Male	Epiphysis clavicle

CHRONOLOGICAL ORDER OF APPEARANCE AND FUSION OF OSSIFICATION CENTERS (*Continued*)

After Birth (Continued)

Age Three	Epiphyses for second and third metatarsals
Female	Epiphyses distal phalanges index and little fingers greater trochanter proximal fibula distal phalanges third and fourth toes
Male	Centers lunate triquetrum epiphysis fifth metacarpal proximal phalanx thumb middle phalanges middle and ring fingers centers tarsal navicular and second cuneiform epiphyses first metatarsal proximal phalanges hallux and fifth toe middle phalanx of second toe
Age Four	Epiphysis fourth metatarsal
Female	Proximal epiphysis radius centers carpal navicular lunate triquetrum multangula minus et majus epiphysis head of fifth tarsal Union of greater tubercle with head of humerus
Male	Epiphyses middle phalanges index and little fingers distal phalanges middle and ring fingers
Age Five	
Female	Epiphyses medial epicondyle distal ulna
Male	Proximal epiphysis radius centers carpal navicular multangulum majus epiphysis distal phalanx little finger greater trochanter center patella epiphyses proximal fibula head of fifth metatarsal middle phalanges third and fourth toes distal phalanges of second third and fourth toes Union of head with greater tubercle of humerus
Age Six	
Female	Epiphysis distal phalanx second toe Union epiphyses middle phalanges third and fourth toes
Male	Distal epiphysis ulna center multangulum minus
Age Seven	
Female	Apophysis of calcaneus
Male	Medial epicondyle Union rami ischium and pubis
Age Eight	
Female	Epiphysis olecranon Union rami of ischium and pubis
Male	Apophysis of calcaneus
Age Nine	
Female	Epiphysis of trochlea
Age Ten	
Female	Center pisiform
Male	Epiphyses trochlea olecranon
Age Eleven	
Female	Epiphysis lateral epicondyle
Male	Center pisiform
Age Twelve	
Male	Epiphysis lateral epicondyle center sesamoid hallux
Age Thirteen	Centers proximal sesamoids thumb
Female	Epiphysis angle of coracoid center sesamoid hallux Fusions angle of coracoid conjoint distal epiphyses to shaft humerus distal phalanx thumb ilium and ischium to pubis distal phalanges second and third toes
Male	Fusions middle phalanges third and fourth toes

Chapter

2

Hereditary, Congenital and Developmental Disturbances

OSTEOGENESIS IMPERFECTA

OSTEOGENESIS imperfecta has been known since the 18th century The disease is also termed fragilitas ossium, idiopathic osteopsathyrosis, periosteal aplasia, and Lobstein s disease The disease involves the tissues developing from the primitive mesenchyme It is a disorder of bone formation characterized by increased fragility of the bones with a varying number of fractures which frequently follow slight trauma The condition occurs at all ages but is seen chiefly in infants and has been diagnosed *in utero* The most common forms of the disease are the nonhereditary, congenital type which appears *in utero* or at birth and the hereditary type In the former, the scleræ may be blue or white and characteristic changes are present in either event The hereditary type may also be associated with blue sclera and atypical roentgen findings The following classification is utilized for completeness and convenience (1) Fetal variety The patient is stillborn or lives only a short time There are numerous fractures of the ribs and long bones The ossification of the skull is incomplete, in many instances the skull being only a membranous bag (2) Infantile variety This is less severe than the fetal type The disease may continue for several years although the prognosis is poor While the ossification of the cranium is incomplete, it is usually much more advanced than in the fetal type (3) Osteogenesis imperfecta tarda or fragilitas ossium The infant is born healthy and may have a normal childhood except for fractures which occur from slight or no apparent cause The fractures may begin at birth or shortly thereafter As the patient grows older the tendency for fractures to occur lessens Blue scleræ deafness of the otosclerotic type peculiar shape of the skull, laxity of the limbs abnormalities of the teeth nails and hair, decreased length of the bones and other anomalies may be present The characteristics of the three types are not always clearly defined

The significant manifestation of osteogenesis imperfecta is the bone fragility which results in an extremely large number of fractures caused by the very slightest possible trauma In the inherited form, the inheritance usually follows a dominant pattern The transmission is generally direct and a parent suffering with the disease transmits the abnormality to about one half of his or her progeny Those that escape the disease do not transmit the condition to their offspring although there are exceptions to this rule There is no sex linkage of the trait There is no preponder-

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FIG 32 Osteogenesis Imperfecta in Childhood *A* Skull at age of one year The bones are markedly thin and porotic with areas which appear uncalcified The anteroposterior diameter of the skull is decreased and there is marked flattening of the lambdoid region *B* The extremities at age of one year The bones are small porotic and bowed There are fractures of the femurs and tibias *C* The extremities at the age of five years The bones are small bowed porotic and present multiple fractures The epiphyses are large The patient was a female and at the age of seven years had a history of forty fractures of the extremities and vertebral column The scleræ were markedly blue

ance of one sex over the other Some of the children of a family who do not inherit the bone fragility may have the blue sclera and brownish coloration of the teeth The change in the sclera is due to deficiency of the fibrous tissue of the sclera which permits the dark pigment of the choroid to shine through The brownish discoloration of the teeth is believed to be due to translucency of the enamel The blue sclera is innocuous However, it is an important characteristic of the disease because a careful review of the literature does not reveal a single case of bone fragility in osteogenesis imperfecta with white sclera The skull resembles an inverted triangle with the base of the triangle representing the top of the skull The forehead is broad and protrudes forward The parietal bones bulge over the external auditory meatuses and the ears are pushed downward and



Fig 31 Osteogenesis Imperfecta Infantile Type There is marked generalized osteoporosis with bowing of the long bones of the lower extremities and multiple fractures

outward The cause of this deformity has never been definitely explained Twenty five per cent of the patients with the disease develop deafness due to otosclerosis This symptom usually appears after twenty-five years of age when the tendency to bone fragility has disappeared or lessened When this point has been reached, the patient often boasts how tough his bones are and that he is now able to do things he formerly could not The etiology of the disease is not known Key believes that the disorder is due to a congenital instability of the mesenchyma or a hereditary hypoplasia of the mesenchyma Some of the signs of the condition can be explained by mesenchymal hypoplasia while others cannot

Roentgen Manifestations The roentgen picture varies according to the type and stage of the disease In the fetal type there is markedly decreased calcification of all the bones There may be numerous fractures of the long bones The ribs and skull are present only as thin, isolated plates of bone The infantile type presents similar but less marked changes In the delayed type, the so called osteogenesis imperfecta tarda, there are many evidences of old united fractures, marked bowing of the long bone and absence of calcification The cortex of the long bones is thin and the medullary cavity markedly widened The shafts of the bones are slender while the extremities are expanded In the healing of the fractures, the extent and distribution of the callus varies widely In some instances there are only very small amounts of new bone formation In others, there is excessive or exuberant callus Bowings and deformities of the bones are common, due to malunion of fractures The cortex is so thin that it is almost invisible At the age of five or six years the shafts of the involved bones become thinner while the width of the epiphysis remains normal This imparts a peculiar elongated appearance to the bones although the actual length of the extremity is normal on measurements The affected bones become markedly bent by malunion of repeated fractures and present bizarre deformities Displacement of the fragment is rare A common finding is subperiosteal fractures with angulation of the fragments In some instances the fractures heal so perfectly that there is no sign of an old fracture except the marked deformity

An interesting phenomenon is lateral displacement of the medullary canal at the site of deformity caused by repeated fractures This is due to the fact that more new bone is laid down on the concave side of the shaft than along the convex surface Narrow, transverse decalcified areas extend into the shaft for a variable distance from the convex side of the shaft and these may grossly simulate incomplete fracture The bony trabeculation is scanty and coarse and the bone has a structureless appearance In later years, the cortex becomes thicker with resultant narrowing of the medullary canal While the upper extremities are involved as frequently as the lower fractures of the upper extremity appear to be less marked There may be fractures of the scapula the ribs, and the small bones The spine is frequently affected In the mild cases there is scoliosis and kyphosis of varying degree and there may be multiple compressions of the vertebral bodies The intervertebral discs are ballooned and may be two or three times the height of the vertebral body There is frequently a marked degree of scoliosis of the lower dorsal and lumbar spine The pelvis is involved and presents a marked deformity with decrease of the lateral diameter due to pressure of the femurs on the acetabulum Because of the low calcium content of the skeleton, a characteristic of the disease is that the roentgenograms appear to be of



Fig 33 Osteogenesis Imperfecta in Childhood *A* Girl 10 years of age Left lower leg The bones are slender and bowed There is marked osteoporosis The left tibia and fibula are fractured and there is marked displacement of the fragments *B* Same patient 15 months later There is a recent fracture of the upper end of the left tibia The bones are slender bowed and markedly porotic *C* Same patient age 12 years There are recent fractures of the bones of the left lower leg and a healed fracture of the lower tibia *D* Age 13 years Left lower leg The porosis bowing and thinning of the bones are more marked than previously The fibula is markedly decreased in size

readily and the healing is characterized by the formation of large masses of cartilaginous callus while bony union is retarded It is believed that the genetic dysfunction of the osteoblasts and odontoblasts is part of a generalized disturbance of the supporting tissue

Laboratory Studies Examination of the blood reveals that the red blood cells and the hemoglobin are normal The white blood cell count varies, there usually being a slight elevation to about 11,000 This may be due to an upper respiratory infection which is frequently present The blood calcium varies between 10 06 and 17 71 mg per 100 cc, with an average of 12 79, indicative of slight elevation The blood phosphorus is within normal limits The alkaline phosphatase is increased in only 30 per cent of the cases

Osteogenesis Imperfecta Diagnosed in Utero The first case in which the diagnosis of osteogenesis imperfecta was established *in utero* was reported by Danelius In a report by Frerking and Zink, a second such case is presented The diagnosis may be established on the basis of the following manifestations (1) poor calcification of the fetal skeleton, especially of the skull and facial bones The fetal skeleton may be entirely invisible on the anteroposterior view, (2) the presence of short, deformed extremities, usually most marked in the lower extremities and (3) the demonstration of multiple fractures It is known that the fetal skeleton can be made invisible by motion of either the fetus or the mother during the exposure If the clinical picture is not clearly understood, the diagnosis of ovarian cyst or a pelvic tumor may be made in error This is particularly apt to be the case if only a single anteroposterior view of the abdomen is utilized Therefore it must be stressed that in all instances in which it is known that a pregnancy is at or near term, failure to demonstrate the fetal skeleton is of the utmost importance and should always lead the observer to consider the possibility of abnormality of the fetus The presence of hydramnios increases the difficulty of visualizing the skeleton particularly if the fetal skeleton is partially or markedly decalcified

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OSTEOPETROSIS OSTEOSCLEROSIS FRAGILIS GENERALISATA

Osteopetrosis or "marble bones is known by the name Albers Schonberg disease It is also termed congenital osteosclerosis and stony bone The condition has been diagnosed on roentgenograms of the fetus *in utero* The disease begins during fetal life and is characterized by obliteration of the medullary spaces marked osseous density and sclerosis, and extreme bony brittleness with multiple fractures There appears to be a failure of resorption of the calcified cartilaginous matrix which normally takes place during enchondral bone development In the skull the involvement is more marked in the bones preformed in cartilage the occipital temporal, sphenoidal, and ethmoidal bones showing extensive changes The bones

very poor quality The sutures and the fontanels are wide The bones of the skull, face, and jaws are markedly decalcified with scattered islands of bone in the calvaria which have the appearance of numerous wormian bones and multiple linear fractures The ossification of the teeth is markedly delayed

Differential Diagnosis In infants and young children the condition must be distinguished from rickets, syphilis and scurvy The normal epiphyseal lines serve to exclude many other conditions The lesion is usually not confused with bone tumors or osteitis fibrosa cystica In osteogenesis imperfecta the blood calcium phosphorus and alkaline phosphatase are usually increased The differential diagnosis from osteomalacia is frequently difficult Osteomalacia occurs in pregnant women, the bones usually bend rather than fracture and the bone trabeculae are made up of osteoid tissue and show a general lack of calcium In osteogenesis imperfecta there is failure of proper evolution of the osteoblasts with defective bone formation Fetal chondrodystrophy and cleidocranial dysostosis may also cause confusion in diagnosis

Pathology There is a difference of opinion as to whether the bones are actually hard or soft Some observers have commented on the extreme density and hardness of the bone stating that it feels and cuts like marble Others have noted that the bones are usually soft This is probably due to the fact that the examinations were made at different stages of the disease the former at about the end stage when the tendency to fragility has been lost and the latter when marked fragility is still present The underlying pathology appears to be deficiency of osteogenesis The epiphyses may be relatively normal until late in the disease when degenerative changes appear Microscopic sections reveal complete loss of the normal bony structure with the Haversian canals appearing as wide spaces interspersed with embryonal and osteoid tissues The bony lamellae are lacking Fibroblasts chondroblasts and transitional cells replace the osteoblasts Sections taken from an area of callus may reveal cartilaginous tissue with extensive areas of necrosis In the diaphyseal portion of the bone necrotic areas are present The condition is nearly always associated with a peculiar developmental disturbance of the teeth termed dentinogenesis imperfecta There is marked, progressive narrowing of the pulp chambers and the canals The abnormality affects only the mesodermal dentin The enamel which is of ectodermal origin is unaffected The disease results from a deficiency of bone tissue both in quantity and quality probably in consequence of a genetic damage to the osteoblasts These appear to be retarded in their function and differentiation so that they form too little bone and at a slow rate and continue to produce primitive coarse fibrillar bone at a time when normal lamellated bone should be developing The osteoclastic activity is not increased and in some instances apparently is reduced

Pathogenesis The bones appear practically normal in length but the growth of the long bones in thickness is more or less impaired This is due to the fact that transverse growth depends entirely on the activity of the osteoblasts while longitudinal growth of bone in the case of the long bones depends primarily on the growth of the epiphyseal and articular cartilage In osteogenesis imperfecta cartilaginous growth and calcification of the cartilage prior to its resorption and replacement by bone are normal The fact that the residual spicules of cartilage are not completely covered by bone does not interfere with the elongation of the bone Fractures heal

readily and the healing is characterized by the formation of large masses of cartilaginous callus while bony union is retarded It is believed that the genetic dysfunction of the osteoblasts and odontoblasts is part of a generalized disturbance of the supporting tissue

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of intramembranous origin the parietals and frontals, are less affected The development of the teeth is delayed The long bones may be involved from one joint surface to the other or only in the central portion of the diaphysis The spine ribs pelvis and other bones of the skeleton are markedly dense and sclerosed as a rule The bone is brittle Pathologic fractures occur with very slight trauma and the patient often is not aware of the presence of a fracture The fractures usually heal The disease is hereditary with recessive genetic features

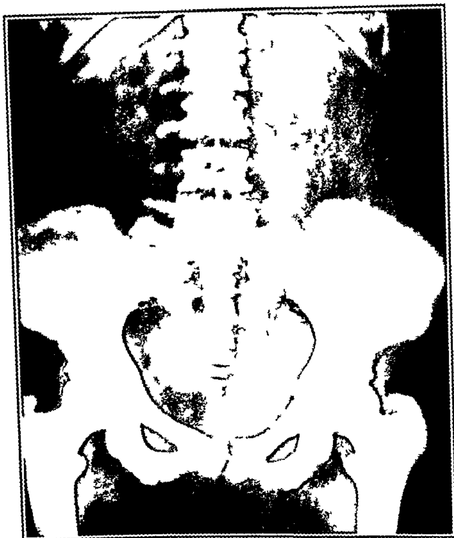


FIG 34 Osteopetrosis The bones are extremely dense and chalky in appearance The normal trabecular pattern is absent The differentiation between the cortex and medulla of the upper femurs is obliterated The changes are generalized and involve practically all of the bones equally

The clinical symptoms of osteopetrosis are manifestations of the encroachment of the bones of the skeleton on the adjacent organs The symptomatology often does not parallel the degree of change in the bones Anemia is usually present and may be of the hyperchromic type although the hypochromic variety is more common The liver spleen and lymph nodes are enlarged Optic atrophy is a frequent occurrence because of narrowing of the optic foramina resulting in poor vision and blindness Pain in the extremities and chest may occur Headaches are a common complaint There is late onset of the menarche and scantiness of men

strual flow, probably due to encroachment of bone on the pituitary body There is usually a familial incidence, and consanguinity of the parents is believed to be an etiologic factor Epiphyseal development and bone growth may be normal indicating that the sclerosis developed after the primary centers of ossification had formed or that the process had not affected the growth of the bones

Roentgen Manifestations There is irregularly increased density and thickness of the bones of practically the entire skeleton The most severe involvement in the skull is at the base, especially the sphenoid bone which shows marked changes in the body and both the greater and lesser wings The sella is normal in size but the floor, the dorsum sellæ, and the anterior and posterior clinoids are thick and dense The temporal bones and occipital bones may also show marked sclerosis The facial bones, the frontal bone, and the parietal bones may not be affected or only to a moderate degree The optic foramina are narrowed and hazy The diploœ is completely obscured and the mastoids and air sinuses are small and undeveloped The suture lines are not obliterated The spine, ribs, clavicles scapulas, pelvis and long bones show narrowing or obliteration of the medullary portions and marked cortical thickening In the tarsal and carpal bones there may be concentric rings of increased density and a small central area of normal bone The iliums present crescentic layers of dense bone laid down over the crests as the bone grows The earliest signs of the disease are frequently in the diaphyses with extension along the shaft The most rapidly growing extremities of the long bones show the most marked changes The pelvis, the vertebræ the skull, the proximal ends of the femurs and the distal end of the tibia and fibula are most severely affected The trabecular structure of the bone is partially or completely obliterated The bones usually remain normal in length A characteristic feature is the presence of transverse and longitudinal bands of increased and diminished density which extend through the metaphysis of the long bones parallel to the epiphyseal line In some instances longitudinal lines are present in the diaphysis parallel to the periosteum

The extent of the changes in the roentgenograms varies widely The whole of a bone including the epiphysis may be uniformly dense and completely without structure The portions which are unaffected may show osteoporosis but this is sometimes apparent rather than real As a rule the metaphyses are more affected than the shafts of the bones The bones are commonly enlarged or clubbed, the enlargement ending abruptly at the junction with the diaphysis Occasionally the reverse is true, the diaphysis being more markedly affected while the metaphyses are less dense and/or even normal or clubbed Except for the clubbing the shape of the bone is not altered In some cases a narrow clear, transverse band marks the junction of the diaphysis and the enlarged metaphysis The carpal and tarsal bones may be universally dense and mottled In other instances they show a dense surface surrounded by a halo of increased radiance The reverse may occur when the pathologic changes begin later in life The phalanges are less commonly affected than the rest of the skeleton and may present a dense transverse band of varying width in the metaphyses in the region adjacent to the epiphyseal line There is decreased resistance to infection and because of the common occurrence of superimposed periodontal and periapical infection the jaws in these patients are particularly susceptible to osteomyelitis Disturbances of dentition may occur in congenital cases or those developing in early post natal life There is

retardation of development and eruption associated with structural defects The roots of the teeth are small and underdeveloped and there is obliteration of the pulp chambers Many of the teeth are missing The bones of the jaw are abnormally dense, particularly the mandible and there is no apparent marrow space in the anterior and premolar regions

Differential Diagnosis The diagnosis is usually easy if the condition is borne in mind Chronic poisoning with lead phosphorus or fluorine must be considered In cretinism juxta epiphyseal bands of increased density may occur These usually disappear under treatment Localized distribution of dense bone may cause confusion with melorheostosis In the early stages during the infantile period the condition must be differentiated from rickets scurvy congenital syphilis excessive vitamin D intake poisoning with heavy metals and acute infantile cortical hyperostosis Hypothyroidism and myosclerosis may produce somewhat similar pictures Syphilis leukemia, tuberculosis Paget s disease Hodgkin s disease and osteoscleroses of the newborn of physiologic origin must also be considered in the differential diagnosis

Etiology The familial occurrence has been so striking that it is believed the disease is carried in certain family groups as a recessive mendelian character There is a high incidence of consanguinity In some instances there is a metabolic condition which resembles chronic hyperparathyroidism in infants It is not known whether this is maternal or fetal in origin It affects both sexes and cases have been described from fetal life to seventy five years of age It is believed that there is some type of faulty calcium and phosphorus metabolism Parathyroid hyperactivity has been thought to play a role in the disease

Pathology The primary cause is thought to be an abnormality of the undifferentiated mesenchymal anlage of the skeleton and bone marrow If the sclerogenous elements of the anlage are principally involved the disease is benign and its clinical course is favorable even though the bone structure is markedly altered On the other hand abnormality of the myelogenous portion of the anlage causes malignant and fatal changes such as commonly occur in infants Blood chemical studies of patients with osteopetrosis reveal normal findings The bones show an altered chemical composition with increased calcium and phosphorus content in all areas and increase in the carbonate in the chondro osseous complex Cohen stresses that the bone lesions constitute the significant morphologic abnormality Hematopoietic deficiencies are the main source of disability although these may bear no qualitative relation to the degree of bone involvement Dietary and infectious influences may be of primary importance in the hematologic picture The transverse lines in the metaphyses of the osteopetrotic bone are fractures in various stages of healing These have presumably occurred because of the increased fragility of the bone particularly at the subepiphyseal region and in the areas where the cortex joins the chondro osseous complex The rapid rate of repair of these fractures contrasts with the delay in remodeling sequences The primary disturbance in the disease is a reduction in osteoclastic activity The thickening of the metaphyses of the long bones is not due to an increased bone production but to a lack of the modeling resorption Similarly the narrowing of the marrow cavity is not a consequence of increased endosteal bone production rather of failure of bone resorption which normally increases the diameter of the marrow cavity The lack of resorption seems to be the consequence of the failure in timing of bone apposition and resorption

Either the apposition of bone is accelerated or its resorption is delayed This disharmony results in a vicious cycle, especially during development and growth of the endochondral bones The growth, degeneration, calcification and initial resorption of the epiphyseal cartilage are not affected The apposition of bone upon the surfaces of the intercolumnar spicules of calcified cartilage proceeds until many of the spaces between the cartilaginous remnants become completely filled It appears probable that the apposition of bone occurs at a normal rate and that resorption which normally removes many of the newly-formed trabeculæ is retarded The consequences of the sclerosis of the bones are twofold Resorption is rendered difficult and the nutrition is reduced

Despite the increased mass of the bone, the sclerotic bone is mechanically inadequate There is no functional arrangement of the bone as this develops only through an alteration of resorption and apposition The sclerotic bone consists not only of bone but also of an irregular mosaic of calcified cartilage and bone The fragility of the bones is the consequence of its structure and not its chemical composition The picture is very similar to that which occurs in fluorosis and syphilitic periostitis The connective tissue in the spaces between the osteophytic trabeculæ may become differentiated into hematopoietic red bone marrow and compensates in part at least for the loss of blood-forming tissue in the marrow cavity Histopathologic study of the bones of the skeleton reveals changes in the epiphyses metaphyses, diaphyses and cortex, which appears to confirm the concept that the pathogenesis is related to disease of the vascular and osteomedullary anlage There is no apparent resemblance of the structure of the bones in osteopetrosis to that of any of several other diseases such as rickets, osteomalacia, syphilis, nonspecific inflammatory disease, phosphorus and fluorine intoxication and osteosclerosis produced by estrogenic hormones

Prognosis Many of the patients die of some process not related to the osteopetrosis In the malignant type death may be early due to intercurrent infection Osteomyelitis of the mandible and maxilla is a frequent cause of death Other causes are anemia and hemorrhage If death does not occur early the children with the disease are often marked for life by hydrocephalus, blindness deafness repeated fractures, and multiple physical abnormalities

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FETAL CHONDRODYSTROPHY—ACHONDROPLASIA

The classical examples of this disease are the king's jester and the circus dwarf with normal sized body, large head, long face, sunken nose, bulging forehead and very short extremities. The condition is due to a cartilaginous dystrophy which is congenital in origin. While it begins in fetal life, the disease may not manifest itself until two or three years after birth. Males are more frequently affected. On histopathologic examination it is found that connective tissue has developed between the epiphysis and the diaphysis. This change may be bilaterally symmetrical or unilateral and

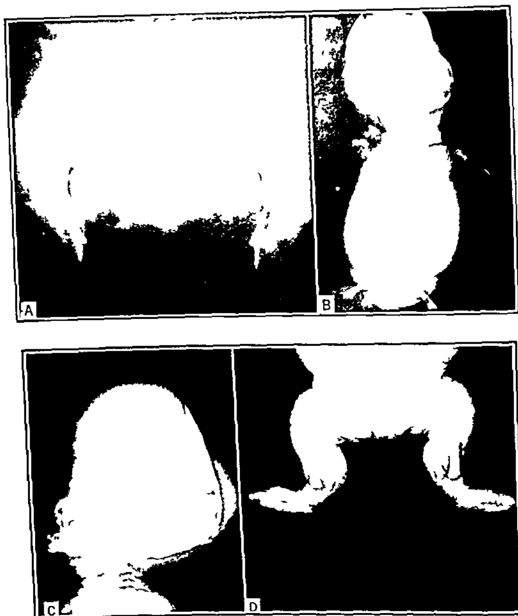


FIG 35 Achondroplasia. Fetal Chondrodystrophy Demonstrated *in Utero*. *A* Roentgenogram of the maternal abdomen during the ninth month of pregnancy. The outline of a markedly abnormal fetal head is visualized in the pelvis. The fetal head is widened and the bones show marked thinning. *B* Roentgenogram of the stillborn fetus. The abnormality of the shape of the head is well visualized. The arms and legs are markedly shortened and the bones show mottling and increased density. *C* Lateral roentgenogram of the skull. There is marked deformation of the skull. The bones of the skull are largely membranous in character with only partial ossification. *D* Roentgenogram of the legs. The bones of the legs and thighs are markedly shortened, bowed and increased in density.

results in retardation of growth of the affected epiphysis In some instances, the centers of ossification of the carpals and metacarpals are delayed in appearing The sphenoid and basilar portions of the occipital bone are preformed in cartilage and disturbance of development of these bones produces deformities of the skull The condition is hereditary in origin and is transmitted as a simple dominant mendelian factor The



Fig 36 Achondroplasia *A* The pelvis and hips The pelvis is small flat and narrow There is coxa vara The lower portion of the acetabulum is absent bilaterally *B* The right femur There is marked bowing and shortening The articular surfaces of the femoral condyles are tilted widened irregular and eburnated *C* Lower legs There is marked bowing The lower legs show less marked changes than the femurs

incidence increases with increase in the age of the mother Many of the patients are stillborn Others survive and have good mentality musculature and sexual development The spine is short and may show kyphosis or lordosis The extremities are short, as are the hands The fingers are of more nearly equal length than normally the middle finger being short The digits diverge and the space between the second and third fingers is wide

Roentgen Manifestations The epiphyseal lines are irregular and oblique, due to involvement of one portion while the remainder develops normally There is irregular expansion of the diaphyses with absence of



Fig 37 Achondroplasia A The chest The deformations and shortening of the humeruses are well shown The bones of the forearm are affected to a lesser extent than the humeruses *B* The pelvis and femurs There is marked deformity of the pelvis and the femurs *C* The lower legs The bones of the lower legs show less marked changes than the femurs and the pelvic bones

the cortex in the affected region Dense, irregular, mottled areas are present in the region of the epiphyseal line Shortening of the long bones is marked and bowing is present in the legs and forearms, the ulna being longer than the radius and the tibia shorter than the fibula The bones of the forearms and legs are less markedly affected than those of the upper arms and the femurs The nose is short and the normal bulge is absent The vertebræ show wedging and irregularity of outline of their margins The pelvis is flat and narrow Coxa vara is common The lower portions of the acetabula may be absent and less than half of the femoral head is in the joint In the skull, there is retardation of the growth, especially of the occiput and sphenoid There may also occur a disturbance of endo chondral bone formation at the basilar synchondrosis Deformity of the skull results with constriction of the base while the vault is large and brachycephalic with increase in the vertical diameter The nose is depressed and sunken the upper jaw receding, the forehead bulging, and the lower jaw protruding The bones of the skull are increased in density The basiocciput is short and the clivus steep The pituitary fossa is small or normal in size The sinuses are large During infancy, hydrocephalus may occur with bulging of the fontanels and encephalographic studies may reveal dilatation of the ventricles

Pathogenesis The condition is characterized by a failure of the epiphyseal and articular cartilage to proliferate by interstitial growth In consequence, there is failure of preparation for the longitudinal growth of long bones and the anterior growth of the base of the skull The principal sites of growth in the cranial base are the intersphenoid and the spheno occipital synchondroses The cartilage in these regions, particularly in the presphenoid and the basisphenoid, under normal conditions disappears in the last months of intrauterine life The cartilage between the sphenoid and the occipital bone continues as a site of growth until the 16th–18th year of life In these synchondroses, the growth of the base of the skull occurs in exactly the same manner as does the growth of the long bones in the epiphyseal cartilages A premature cessation of cartilaginous growth with consequent premature synostosis of the bones of the base of the skull results in permanent cessation of growth in this region and is responsible for the lack of facial growth in depth as the bones of the upper portion of the face are attached to the skull Since the brain in these dwarfs grows normally in volume, it is forced to change its shape and the bones of the vault of the skull undergo marked bulging The bulging of the forehead increases the impression of a deeply sunken and flat upper face The condyloid cartilage normally grows both interstitially and by apposition whereas the epiphyseal cartilage grows in the long axis of the bone by interstitial growth only In the chondrodystrophic dwarf the interstitial proliferation of cartilage is arrested, but not its appositional growth, and the epiphyseal cartilages cease to grow altogether Under these conditions the cartilage in the condyle of the mandible can continue its growth because the appositional growth of cartilage compensates in part at least for the lack of interstitial proliferation The epiphyses of some of the bones, particularly the head of the femur and the humerus present a mushroom shape The widening and flattening of the bony ends is a consequence of the dual growth mechanism of the articular cartilage The growth in thickness is entirely interstitial The growth in area is accomplished by appositional growth at the borders and these are covered by an extension of the connective tissue of the articular capsule Arrest of

interstitial growth prevents lengthening of the epiphyses, which however continue to expand at their peripheries. The formation of a rosary at the ends of the ribs at the point where they join the costal cartilages is explained in a similar manner.

There is marked disproportion between the short extremities and a fairly normal trunk probably due to the great number of centers of growth in the intervertebral discs which contribute to the longitudinal growth of the vertebral column and their stimulation by the growing viscera. There is marked change in the pelvis and this is particularly important during pregnancy. There are 2 types of pelvic deformity. In one the pelvis is flat anteroposteriorly. The entrance into the pelvis is kidney shaped and the sacrum occupies practically a horizontal position. In the second type there is a generalized narrowing without marked deviation from the normal shape. Because of the retardation of bone production at the epiphyseal centers while periosteal osteogenesis continues in an undisturbed fashion, bone growth is diminished in the longitudinal direction while circumferential growth proceeds as usual. As a result the long bones become short and thick. There is frequently interference with dental development and eruption. This is manifested by a tendency to retarded eruption and noneruption of the teeth. The presence of many unerupted teeth presents a picture which is similar to that in cleidocranial dysostosis.

MULTIPLE CARTILAGINOUS EXOSTOSES HEREDITARY MULTIPLE EXOSTOSES DIAPHYSEAL ACLASIS HEREDITARY DEFORMING CHONDRODYSPLASIA

One of the most common of the hereditary malformations of the skeleton is the so called multiple cartilaginous exostoses. As the malformation has its origin in a disorder of growth of the skeletal cartilage the condition has also been termed hereditary deforming chondrodysplasia and diaphyseal aclasis. The disease is not manifested in the infant but develops gradually during childhood. It is more common in males than in females. The genetic mechanism has not been fully determined and it can only be stated at present that the etiologic factor is dominant and incompletely sex linked. The condition is characterized by the gradual development and growth of many knobby protuberances on different bones. The number of exostoses is widely variable and may reach large numbers cases with several hundred having been reported. The most common sites are the metaphyseal regions of the bones of the knee ankle shoulder scapula pelvis ribs vertebræ metacarpals and metatarsals. The bones of the skull are seldom involved. The excrescences are rarely unilateral the lesions usually being symmetrical. The growth of the exostoses parallels that of the skeleton and the exostoses cease to grow with cessation of growth of the skeleton. If one or more of the exostoses continues to grow after this period it is essential to consider the diagnosis of sarcomatous degeneration. In severe cases the growth of the skeleton is impaired particularly the long bones. In consequence the shortness of the limbs causes a resemblance to the proportions in chondrodystrophy. An important factor is the frequent occurrence of bowing of one or both bones of the forearm and the leg. This may result in disarticulation of the forearm and hand with consequent severe limitation or restriction of motion

The exostoses occur most commonly in the diaphyses of the long bones In the ribs they are usually limited to the bony areas The epiphyses of the long bones are never involved This indicates that the exostoses develop in the growth zone of the cartilages adjacent to the metaphyseal region of the bone and that they shift during growth to the diaphysis

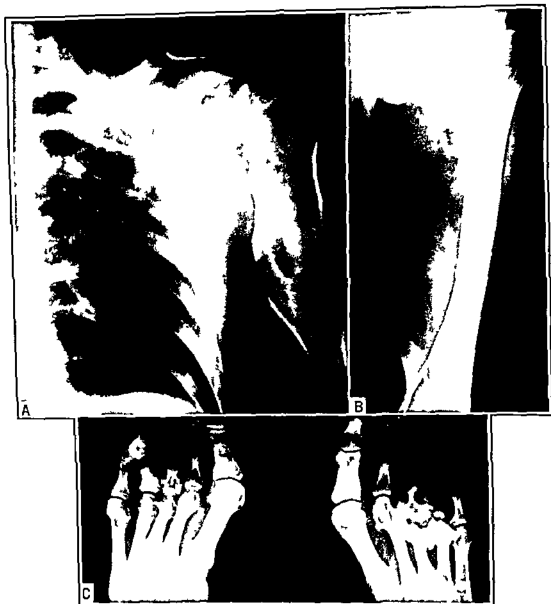


FIG 38 Multiple Cartilaginous Exostoses *A* Right shoulder There are multiple exostoses in the upper third of the humerus the superior aspect of the scapula and the first and third ribs *B* Left femur Exostoses are present at the superior and inferior aspects of the femur *C* Both feet There are multiple exostoses bilaterally with growth disturbances of the third and fourth toes The changes are more marked on the right side

The exostoses are made up of cartilage which undergoes destruction and replacement by bone from its deep surface while the superficial layers continue to grow In the later stages, the exostosis is composed of a base or stalk and a core of spongy bone with a covering of hyaline cartilage The marrow spaces of the exostosis communicate with those of the bone which carry the exostosis Because of this fact the exostosis cannot be regarded as a growth upon the bone but must be considered as part of a

disfigured bone When normal growth has terminated the cartilaginous covering of the exostosis disappears and is replaced by bone The exostoses are covered by a layer of dense connective tissue which comprises a continuation of the normal periosteum If a tendon slides over an exostosis, a synovial bursa may develop

Pathogenesis The exact mechanism by which the genetically determined exostoses develop has not been established Most of the exostoses and in many instances all of them, develop from epiphyseal cartilage or from cartilage which serves an analogous function in the growth of the bone The exostoses are not caused by an abnormal manifestation of the dormant potency of the periosteum to produce cartilage It has been



Fig 39 Multiple Cartilaginous Exostoses A Left knee There are exostoses at the lower end of the femur and the upper ends of both bones of the lower leg *B* Left forearm There is impairment of growth of the lower end of the ulna with resultant deformations of both bones of the forearm The proximal end of the radius is anomalous and is disarticulated The ulna is stunted in growth while the radius is relatively elongated

stated by some observers that the abnormal proliferation of the juxtametaphyseal cartilage is caused by lack of a bony covering which normally is provided by the extension of the periosteal cuff of the diaphysis over the cartilage Although the perichondral or periosteal bony collar in many instances extends beyond the plane of the metaphysis at the periphery of the cartilage this extension is not always possible In the older stages of osseous development the juxtametaphyseal cartilage is usually covered by perichondrium rather than by bone It is believed that the exostoses are a distortion of the normal growth pattern of the bone More specifically, it is considered a distortion or disharmony in the pattern of the growth of the cartilage the proliferation of which produces not only the enlargement but also the primary modelling of the bone The growth of epiphyseal cartilage is interstitial in the longitudinal direction while it is

appositional in the transverse direction A harmony between these factors is essential for normal development and a disturbance of the harmony with predominance of appositional growth results in distortion of the bony outline with irregular thickening in the lateral direction If the disturbance of the growth pattern of the cartilage is severe, there will not only be an increase in localized transverse growth, but also a decrease in longitudinal growth If this imbalance of proliferation is localized to a certain area of the circumference of the epiphyseal plate, the deficiency of longitudinal growth may be restricted to a part of the cartilage while other segments continue to proliferate normally This uneven longitudinal growth results in a curving of the involved bone In the forearm, the ulna is often stunted in growth while the radius is curved and relatively elongated, producing the so called Madelung's deformity Normally these bones grow at the wrist at an equal rate It appears that there is a different transverse diameter in the radial and ulnar epiphyseal plate A narrower plate will be most apt to be stunted in its longitudinal growth as a whole while unequal growth may be expected in a relatively wide epiphyseal plate The transverse diameters of the epiphyseal plate at the ulnar head are about one-third to two-fifths those in the distal radial epiphyseal plate

Because of the failure on the part of the periosteum to model the metaphyses of the long bones in the normal manner, Keith suggested the name diaphyseal aclasis He stated that the bones formed entirely within cartilage or entirely within membrane including the tarsal and carpal bones the epiphyses of the long bones, the sternum, the bodies of the vertebræ, the bones of the cranial vault and the face are not affected in the disorder, the condition involving only those parts of the skeleton in which bone laid down in cartilage becomes covered with periosteal bone He considered it an arrest of development in the periosteal ring which permits the cartilage of the growth disc to be exposed on its surface and free to form exostoses Other authors have found that bones formed strictly in cartilage or in membrane are not invariably spared as stated by Keith but are occasionally the site of an exostosis

Differential Diagnosis Diaphyseal aclasis may resemble dyschondroplasia (Ollier s disease) since exostoses on the surfaces of the metaphysis may produce irregular mottling which suggests cavitation of the bone The masses of calcified cartilage seen in some instances of diaphyseal aclasis are not endosteal as in dyschondroplasia They are caused by local disturbances of endochondral ossification in the proliferating cap of hyaline cartilage overlying the exostosis Small exostoses may arise near the epiphyses of the long bones in dyschondroplasia but are rough, irregular, point in any direction do not grow larger and are in many respects unlike those in diaphyseal aclasis In dyschondroplasia, expansion of the ends of the long bones is very seldom seen, while this is a prominent feature in diaphyseal aclasis Dyschondroplasia and diaphyseal aclasis are not related and must be considered separate and distinct entities with different roentgenographic manifestations

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DYSCHONDROPLASIA OLLIER'S DISEASE

Ollier described a syndrome closely allied to the chondromas which he termed dyschondroplasia The condition is a developmental anomaly of endochondral ossification rather than a true neoplasm and is characterized by an accumulation of hyaline cartilage in the ends of the shafts of the long bones, more commonly in the lower extremities than the upper There is shortening due to interference with longitudinal growth Dwarfism may result If the accumulation of hyaline cartilage is more marked on one side than the other there is greater interference with growth on the side of the more marked involvement with resultant angulation and deformation of the bone There may be slight expansion of the cortex

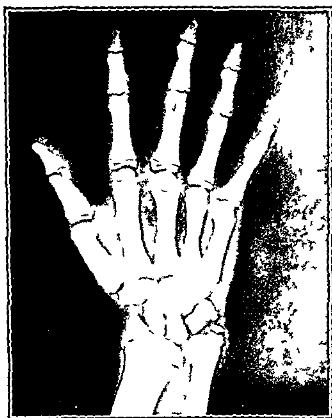


Fig 40 Ollier's Disease Dyschondroplasia Right hand There are multiple enchondromas of the first and second metacarpals the proximal phalanx of the thumb and the middle phalanx of the index finger

The dyschondroplasia *of Ollier's disease is characterized* by a disturbance of bone formation in cartilage particularly at the growing ends of the bone The cartilage persists as islands in the diaphysis and undergoes proliferation to form enchondromas In some situations particularly in the phalanges *there may be great expansion and deformity* of the bone The enchondromas are formed particularly near the epiphyseal lines Because of the imperfect ossification and irregular growth there is bowing of the affected bones which may lead to marked asymmetry of the skeleton The stresses of *weight bearing often cause secondary* deformities The patients are usually small in stature and show marked muscular atrophy because of the delay in skeletal growth While the lesions may affect any portion of the skeleton involvement of the long bones and phalanges is

common and of the skull, carpus and tarsus uncommon Fractures occur readily and often are slow to unite Malignant changes may develop in the chondromas In some cases there are severe deformities requiring amputation In the prepuberal years, small nodules about 1–2 cm in diameter appear on the small bones of the hands and feet, to be followed by others, some in the long bones The distribution is unilateral or markedly asymmetrical Fractures may follow trivial injuries The development of the two sides of the body may be unequal and bony deformities such as pes planum, genu valgum, and scoliosis are present with marked deformations of the bony structures The alterations in the hands and feet attain grotesque proportions in severe forms of the disease The

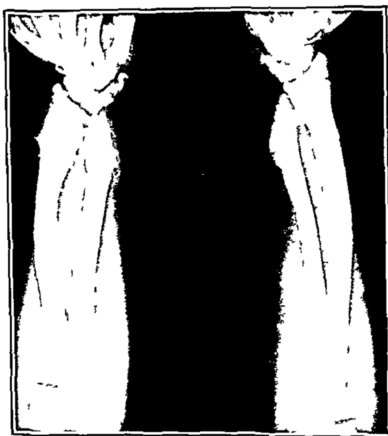


Fig 41 Ollier s Disease Dyschondroplasia with Madelung s Deformity There are enchondromas of both bones of the forearms The interference with growth of the bones has produced the characteristic Madelung s deformity of both arms

condition becomes stationary after full growth has been reached The intelligence is usually average

The roentgen changes are striking The small bones of the hands and feet exclusive of the carpus and tarsus present multiple cystic areas the enchondromata with irregular expansion of the cortex and widening of the shaft resulting in marked irregularity of contour of the bones Similar changes are present in the long bones and less commonly in the ribs scapulae and vertebræ The roentgen appearances vary with the stage of the disease In the initial phases there are lines of condensation in the spongiosa During the period of evolution, the process spreads from the metaphysis into the diaphysis The primarily endosteal process extends through the cortex to cause changes in the external contour of the bone Large cyst formations are common The stage of involution is character-

ized by calcification of the cartilaginous masses and periosteal bone formation There is a tendency to spontaneous repair Malignant degeneration is characterized by rapid growth of the enchondromas When the disease involves the long bones, there is apt to be shortening and bowing When one of a pair of bones is affected, as in the case of the forearm, the so called Madelung's deformity may occur

MAFFUCCI'S SYNDROME DYSCHONDROPLASIA WITH HEMANGIOMAS

Maffucci in 1881 described an entity characterized by dyschondroplasia and multiple hemangiomas It is a rare condition only twenty seven cases having been reported in the literature since that time The disease is non familial and the patient is usually normal at birth Involvement is unilateral or extremely asymmetrical, there being uneven growth of the two sides of the body The disease is more common in males The onset is prior to puberty There is an associated susceptibility to fractures and phleboliths are a common occurrence in the hemangiomas Chondrosarcoma apparently develops in approximately 20 per cent of the cases and for this reason the prognosis is less favorable in Maffucci s syndrome than in dyschondroplasia alone In a case recorded by Mullins calcification was demonstrable medial to the upper third of the humerus in an area which corresponded to the location of a superficial tumefaction There was reduction of the size of the right humerus and the bones of the right forearm as compared with the corresponding areas on the left There were fewer epiphyses on the right in the patient at the age of six There was slight reduction in the size of the left hemipelvis and the soft tissues of the buttocks The left femur was longer than the right and showed decided bowing

Maffucci s syndrome has two distinct components the dyschondroplasia being identical with Ollier s disease and the vascular abnormalities consisting of cavernous hemangiomas and phlebectasia The hemangiomas may occur anywhere in the subcutaneous tissues and have also been noted in the lips, the palate and the mesocolon They form red or reddish blue tumors which are soft compressible and sometimes tender on pressure Thrombi may develop and sometimes become calcified When phleboliths are present they produce a striking radiographic picture The phlebectasia is common and may affect large groups of veins or be confined to a few local areas with resultant bead like swellings The condition appears to originate from a dysplasia of the mesoderm There is nothing to suggest that either of the component abnormalities induces the other The subjects are normal at birth and the lesions are first noted in most instances during childhood The condition progresses and causes increasing disability but may become static after the growth period has ended However the incidence of malignant degeneration in the chondromas is high Involvement of the skull is uncommon but cases have been reported with chondrosarcoma of the body of the sphenoid When the lesion involves the skull the clinical picture may be that of an intracranial neoplasm There may be subarachnoid hemorrhage due to hemorrhage into the tumor

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CHONDROECTODERMAL DYSPLASIA ELLIS-VAN CREVELD DISEASE

Ellis and Van Creveld in 1940 described the syndrome of chondro ectodermal dysplasia The condition is characterized by dysplasia of the cutaneous appendages with anomalies of the hair, teeth and nails, polydactylism involving the hands, and dwarfism due to shortening of the legs Caffey has described the findings in three additional cases The changes may vary somewhat in different patients and in the various age groups However, the condition is a definite congenital syndrome which must be differentiated from the numerous other congenital dysplasias and the various forms of dwarfism The teeth and nails have been affected in all of the recorded cases Both the upper and lower teeth are defective and show retardation of development The nails of the hands and feet are small, abnormal in shape and friable In the presence of supernumerary digits, the nails of the rudimentary sixth digits are absent The hair may be normal in amount or alopecia may occur The upper lip may be fused with the gum pad Congenital heart lesions occur in many instances with left sided hypertrophy and harsh systolic murmurs, most probably due to the presence of patent interventricular septum

The tubular bones of the extremities are short and thick, the shortening being absolute and in proportion to the trunk Dwarfism is due to shortening of the legs Caffey states that the pattern of the alteration of the bones of the extremities, the progressively increased shortening from the trunk to the distal phalanges, constitutes a newly recognized and diagnostic feature of the disease The root bones of the extremities, the femurs and humeruses, show the least change while the middle and distal phalanges are shortened the most In some cases the primary ossification centers of the distal phalanges fail to appear In chondroecto dermal dysplasia the femurs and humeruses are relatively the longest, while in achondroplasia, these bones are relatively the most markedly shortened The fibulas are shortened in chondroectodermal dysplasia while in achondroplasia they are relatively the longest

There are characteristic deformities of the bones in chondroectodermal dysplasia with enlargement of the proximal ends of the ulnas and the distal ends of the radiuses the distal ends of the ulnas and the proximal ends of the radiuses being decreased in size There is associated dislocation of the radial heads from the elbows in many cases An important manifestation of the Ellis Van Creveld syndrome is widening of the proximal ends of the tibial shafts with long lateral slopes and short medial slopes The proximal tibial epiphyseal cartilages are associated with thin hypoplastic ossification centers In the hands there is a rudimentary sixth digit at the ulnar aspect There are six metacarpals the fifth and sixth metacarpals showing side to side fusion the unfused aspect of the fifth metacarpal being markedly concave The capitate and hamate bones are fused The feet rarely show polydactylism and no fusions occur in the metatarsals or tarsals There is retardation of maturation of the primary ossification centers of the phalanges and acceleration of maturation of their secondary ossification centers in the epiphyseal cartilages The sesamoids of the hands show retardation of maturation

Dental aplasia may vary from absence of a limited number of the permanent teeth to total absence of both the primary and permanent teeth Cases of ectodermal dysplasia in which there has been total anodontia have

been reported The degree of dental aplasia is dependent upon the time of onset of the ectodermal disturbance Cases which have their onset early in fetal life are characterized by total aplasia With later onset of the condition, the primary teeth may remain unaffected while all or a few of the permanent *teeth may fail to develop The teeth which are most* apt to remain unaffected are those for which the anlage is first laid down, that is the incisors, canines, and first molars The jaws are small and underdeveloped, particularly the mandible The roentgen appearance is similar to that in the senile edentulous jaw in which the alveolar process has undergone resorption after loss of the teeth In instances of total anodontia, there is no need for the alveolar process and it fails to develop

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MORQUIO S DISEASE OSTEOCHONDRODYSTROPHY DEFORMANS

Morquio s disease is a hereditary dystrophy of cartilage and bone which results in dwarfism and generalized skeletal deformity It tends to run in families The following characteristics are present in the disease There is marked spinal kyphosis with angulation at the dorsolumbar region The chest is deformed with an increase in the anteroposterior diameter and the sternum is thrust forward horizontally to a point The clavicles are raised and the neck is short There is genu valgum due to enlargement of the internal femoral condyles The feet are large and flat with varus deformities There is enlargement of the head and trochlea of the humerus There is laxity of the ligaments low muscle tone grating and crepitation of the joints and difficulty in locomotion In some instances there is a lowering of the serum calcium level The other biochemical findings are normal

Roentgen Manifestations One of the most important findings is irregularity and fragmentation of the epiphyses with an appearance of multiple centers of ossification The long bones are short and thick with coarse irregular reticulation of the cancellous tissues and absence of regular lines in the lamellae There is platyspondylia the vertebrae being wide and flat some being pointed in front and others wedge shaped The intervertebral discs are wide The carpal and tarsal bones are irregular and the bases of the metacarpals and metatarsals are pointed The prominence of the thorax is due to arrest of growth of the spine at the cervicodorsal junction The epiphyses of the long bones are irregular enlarged flattened and of varying density suggesting localized destruction The femoral head may be absent misshapen or fragmented The acetabulum and the glenoid fossa may be irregular The diaphyses are thinned and show areas of rarefaction near the ends The cortex of the long bones may be thickened on the side of greatest strain The small bones of the hands and feet are shortened and show thinning of the cortex irregularities of the epiphyses and increased trabeculation The ossification centers for the carpals and



FIG 4? Osteochondrodystrophy Deformans Morquio's Disease *A* Dorsolumbar spine There is marked kyphosis in the dorsolumbar region with wedging of the upper lumbar vertebrae *B* Right arm The bones of the arm are short deformed and abnormal in architecture *C* The pelvis and femurs The pelvis is asymmetrical wide and flat The hips are deformed *D* The lower legs The bones are short broad and bowed The articular surfaces are wide and flat

tarsals are retarded There is kyphosis in the dorsal and lumbar areas The ribs are wide and flat and the intercostal spaces are narrowed There is anterior protrusion of the sternum The long bones show no unusual fragility The joint spaces, especially the shoulder, wrist knee hip and ankle, are widened and show overgrowth of cartilage The epiphyses of the larger joints, particularly the hips, undergo deterioration The ossific nuclei which originally were well formed disappear and no further ossification occurs The muscles become progressively weaker with the result that the joints are unstable and dislocation of the hips occurs The acetabulums expand and the upper ends of the femurs develop a valgus deformity and come into apposition with the lateral aspects of the iliums Later this contact is lost and the joints become flail in type The opposing bony surfaces are irregular in outline and are separated by dystrophic cartilage The knee joints develop a knock knee deformity without bowing of the shafts of the diaphyses as in rickets In the forearm the dystrophic changes are associated with bowing due to irregularity of growth

Etiology The condition occurs in both sexes There is frequently consanguinity of the parents Syphilis has not been found in these patients It is believed to be due to a hereditary factor and all cases appear to have an associated constitutional weakness

Differential Diagnosis Florid rickets in infancy is characterized by generalized osteoporosis and the outlines of the diaphyseal extremities and *the ossific nuclei for the epiphyses are blurred* The epiphyses ossify from one central nucleus The bones bend with weight bearing and the skeletal deformities are symmetrical and generalized The bones fracture easily and multiple pseudofractures may develop The condition responds favorably to adequate dosage of vitamin D with arrest of the deformity and cure in many instances In hypothyroidism there is generalized delay in ossification and the epiphyses are late in developing ossific nuclei When these do appear they are multiple and with weight bearing and normal function they become deformed or unstable The enlarged epiphyses particularly in the femoral capital epiphysis show these changes to the greatest extent The disease is cured by adequate thyroid medication In osteochondritis usually a single epiphysis is affected although the changes may be multiple In some instances the upper femoral epiphyses are involved bilaterally In these cases the extent of the involvement is not the same on both sides, indicating that the process started at different times Deformities are due to pressure during the plastic stage of the disease In achondroplasia the long bones and the bones of the base of the skull are prematurely fused shortened and thickened and the bones of the pelvis are wide The vertebral bodies tend to be square in shape and at birth there is a localized dorsolumbar kyphosis without change in the form of the bodies The epiphyseal growth is from single nuclei and progresses regularly to maturity The constitution is good and the musculature is well developed whereas in chondro osteodystrophy the defect appears to be due to faulty building material The two conditions achondroplasia and chondro osteodystrophy may occur together Patients with gargoylism may exhibit generalized irregularities in the growth of the skeleton which produce dwarfism They also may have defective development of other systems such as the nervous circulatory respiratory alimentary and endocrine

DYSOSTOSIS MULTIPLEX LIPOCHONDRODYSTROPHY

The syndrome termed gargoylism is known under a variety of names, the chief of which are dysostosis multiplex, chondro osteodystrophy, and lipochondrodystrophy The names of Thompson, Hunter, Hurler, and Pfaundler in various combinations have also been applied to the condition, but in conformity with the modern practice of avoidance of the use of proper names to characterize disease processes, the names are mentioned merely for historical purposes Thompson observed the disease in 1900 The condition was described by Hunter in 1917, by Hurler in 1919, and by Pfaundler in 1920 Gargoylism is a congenital familial syndrome which produces striking skeletal changes The condition is rare and only a relatively small number of cases, probably less than 200, have been reported in the literature

Etiology The cause of the condition is unknown The existence of a genetic factor appears probable as there is a high incidence in siblings and also of marriage of cousins among the parents of affected children The inheritance appears to be through a single gene The skeletal changes are a form of chondrodystrophy and the condition is considered a manifestation of a defect in the germ plasm There is a strong familial tendency The disease is believed by some to be due to the intracellular storage of fat in the central nervous system and of glycogen and glycoprotein throughout the body, particularly in the bones

Pathology The term gargoylism is merely descriptive of the general appearance of the patient The term lipochondrodystrophy is more significant and indicates a possible association with the lipoidoses of Hand Christian Schuller s Gaucher s, and Niemann Pick s disease The important histopathologic feature is a degenerative change in the nerve cells in the central nervous system There are cytoplasmic lipoid deposits in the basal ganglia and the brain stem Perivascular extracellular lipoid is present in the gray matter The lipoid is considered a cerebroside There is no lipoid infiltration in the bones but it has been identified in the liver spleen, lymph nodes, lung heart and sinuses The basis of the changes in the skeleton is thought to be the escape of cerebrospinal fluid through a gap in the roof of the developing fourth ventricle which results in the formation of blebs in the subcutis of the embryo Failure of absorption of these blebs may produce all of the defects which have been described It has also been stated that the disturbed lipoid metabolism is due to the effect of the bleb formations on the pituitary and the hypothalamus Abnormal granulation of the lymphocytes has been described In some instances there has been found hydrocephalus with atrophy of the temporal lobes

Clinical Manifestations Many or all of the following clinical manifestations occur The head is large and the forehead prominent The face is broad with coarse, ugly features and the neck is short The nasal bridge is flat and wide the nostrils are big, and the lips are thick and partly open due to a large protruding deeply fissured tongue Dentition may be delayed and the teeth are widely spaced and poorly developed The patient presents a grotesque appearance The palpebral fissures are narrow There is mental retardation and clouding of the corneas in some instances The abdomen is protuberant and the liver and spleen are large and easily palpable There is usually an umbilical or an inguinal hernia The skin is puffy The appearance of the extremities is striking The patients have short legs and arms the joints are held in semiflexion and complete extension

is usually not possible The hands are broad, short, and claw like, the fingers being partially flexed An important manifestation is a severe lumbodorsal kyphosis Other less constant clinical features are muco purulent rhinorrhea, recurrent otorrhea deafness, underdevelopment of the muscles hypertrichosis retarded sexual development and a coarse dry skin Dwarfism is a prominent feature in later life, although the growth of the patient during the first years of life is not affected The number and degree of the various components are widely variable

Roentgen Manifestations On roentgen examination, it is found that abnormalities are present throughout the entire skeleton The skull is large, frequently with thick walls There is protrusion of the forehead and supra orbital ridges and a prominent ridge may be present in the mid line at the sagittal suture The skull may be scaphocephalic brachycephalic, or oxycephalic The sella is large and deep There is hypertelorism There may be prominence of the convolutional markings of the skull The maxilla and mandibular rami are underdeveloped and the angles of the mandibles are increased A kyphosis is present in the lower dorsal and upper lumbar region and the vertebrae in this region may be wedged The supero anterior aspect of the vertebral body is deficient while the infero anterior border forms a hook like extension The scapulas are widened The ribs present narrow roots broad bodies and distorted necks often leaving the spine at right angles The clavicles are wide and deformed

The osseous changes are more marked in the upper than in the lower extremities There is retardation of epiphyseal maturation abnormal configurations of the shafts and irregular cartilaginous proliferation at the epiphyses The shafts are short and stubby and exhibit bizarre swellings in the central portions with taperings toward the ends This latter feature is a significant diagnostic sign The head of the humerus is short and exhibits a valgus angulation The articular surfaces and the epiphyseal plates of the lower ends of the radius and the ulna are tilted toward each other The metacarpal bones and the proximal portions of the metatarsals and the phalanges are broad and shortened The first two phalangeal bones are pointed at their ends Both acetabulums are shallow and the ischiums are disproportionately small in relation to the iliums The femurs present a coxa valga angulation of the neck and the head is flat and malformed The femoral necks are broad Genu valgum is invariably present due to the tilted epiphyses and abnormal lines of the femurs and tibias The feet show the same characteristics as the hands The epiphyseal plates of the long bones are flat and deformed and present irregular outlines of ossification Retarded epiphyseal age is shown by about half of the patients

When fully developed the roentgen manifestations are characteristic The bones of the arms hands and ribs show the most striking changes the legs being little affected The cardinal lesion in the tubular bones comprises swelling of the shaft due to distention of the medullary cavity The cortical walls may be thickened in the early years of life but are usually thinned Uneven increase in the girth of the shafts is the most important single diagnostic sign and without this manifestation the diagnosis cannot be established with certainty There is incomplete modelling and tubulation of the long bones with resultant deformation of the external contours of the bones In some instances the ends are swollen and widened and the central segment is narrowed producing a dumbbell formation The changes at the cartilage shaft junction and the epiphyseal plates are slight and do not aid

in the diagnosis The paucity of metaphyseal changes associated with the marked diaphyseal expansions differentiates gargoylism from the osteochondrodystrophies In the earliest stages of gargoylism as seen in early infancy, the roentgen changes differ from those in later life The early manifestations comprise generalized rarefaction of the bones, cupping and spreading and may strongly suggest rickets or hyperparathyroidism

Differential Diagnosis In the fully developed stage of gargoylism, the picture is characteristic and the diagnosis is established on the presence of the following groups of symptoms (1) Epimetaphyseal dysostosis, short growth, stunted trunk, lumbar kyphosis and deformities of the sternal bone, short and sometimes distorted extremities, short, broad and stubby fingers, the head is large and may be scaphocephalic with prominence of the forehead and bulging of the parietal bones, reduced mobility of the articulations resulting in a claw hand, or in some cases, increased mobility of the joints (2) The manifestations of lipoid or, according to some authors, glucoprotein storage with enlargement of the liver and spleen, cloudiness of the corneas, progressive mental deterioration, and abnormal granulations in the white cells and the bone marrow (3) A group of accessory signs which comprise the gruesome gargoyle facies, hypertrichosis, flabbiness of the skin, umbilical hernia, and a tendency toward frequent upper respiratory infections The most important condition which must be considered in differential diagnosis is Morquio's disease In Morquio's disease, the skull and sella turcica are normal whereas in gargoylism it is enlarged and has a shoe shape In Morquio's disease the vertebral bodies show platyspondyly with irregularity of the epiphyseal plates In gargoylism the vertebral bodies have a beak-like protrusion of the anterior edges particularly marked in the second and third lumbar vertebræ In Morquio's disease, the long bones have normal diaphyses, while in gargoylism the diaphyses are enlarged and taper toward the metaphyses In Morquio's disease, the epiphyses are small irregular ill defined and fragmented, while in gargoylism they are bigger In Morquio's disease, normal or delayed bone maturation is found in the carpal ossification centers and the ribs are normal In gargoylism there is delayed maturation of the carpal ossification centers and the ribs present a spatulated form or appear like oars Many cases of Morquio s disease present a superficial resemblance to gargoylism yet lack one or more of the cardinal features of the syndrome such as the corneal clouding or mental retardation These have been termed "formes frustes and form an intermediate condition between gargoylism and Morquio s disease Also the abnormalities are usually not noted until walking is attempted while in gargoylism the child is noticed to be abnormal at birth because of the characteristic facies The joints in Morquio s disease are lax and motion is not restricted as in gargoylism Most cases of the former condition have a coxa vara type of deformity in contrast to the coxa valga of gargoylism There is more involvement of the vertebræ in Morquio's disease, most of the spine being affected while in gargoylism only a few vertebral bodies are involved in the chondrodystrophic process

Severe rickets causes a disproportionate dwarfism but the deformities are more marked in the lower extremities due to weight bearing The head is more square in shape and shows frontal bossing The epiphyseal lines fuse late rather than early as in gargoylism In rickets, the joints are hyperflexible and there is no mental retardation or corneal clouding

Cretinism must be considered in differential diagnosis Thyroid therapy has been used in many cases of gargoylism without improvement This is important in differential diagnosis as is also the absence of corneal opacity enlargement of the liver and spleen and mental retardation Syphilis has been suggested as a possible diagnosis However the bone changes are quite different and the Wassermann test is negative Hypophyseal dwarfs present slender bones without deformity and also underdevelopment of the genitalia and the secondary sex characteristics In the early stages the manifestations are similar to those in Leri s disease and the diagnosis cannot be established by roentgen methods alone clinical studies being essential to reach a final conclusion

ADDITIONAL READING

HUNTER C Rare Disease in Two Brothers Proceedings Royal Society Med *10* 104 1916–1917
HURLER G Ueber Einen Type Multipler Abartungen Vorwiegend am Skelett System Ztschr F Kindrh *24* 220 1920
HARVEY R M Hurler Pfaundler Syndrome (Gargoylism) A Review of the Literature with the Report of a Single Case Am J Roent *48* 732 1942
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LERI TYPE OF POLYTOPIC ENDOCHONDRAL DYSOSTOSIS

In the Leri type of polytopic endochondral dysostosis the usual age of onset is about two or three years The head and facies are usually not remarkable although saddlenose may occur The growth may be stunted with varying degrees of dwarfism There is little or no mental retardation The neck may be shortened and slight or severe deformities of the trunk with varying degrees of lordosis occur The arms and hands are short The diaphyses of the long bones are increased in density and thickness and show bowing Wedge shaped deformities of the vertebræ and hemivertebræ have been described The changes are not characteristic and the diagnosis can be established only by correlation of the family history and the hereditary background together with the clinical and roentgen manifestations

CLEIDOCRANIAL DYSOSTOSIS OSTEODENTAL DYSPLASIA

Cleidocranial dysostosis was first established as a clinical and pathological entity in 1898 by Marie and Sainton They stressed the cardinal features of the disease which included the hereditary transmission hypoplasia of the clavicles increase of the transverse diameter of the skull and delay in ossification of the fontanels In 1899 Terry added certain important pathognomonic changes His patients showed faulty eruption of the teeth scoliosis and poor ossification of the pubic and ischial bones In the previously reported cases there is a proportion of about 8 to 5 in favor of a familial tendency Transmission appears to occur equally through the male and female The sex distribution has been approximately equal The anomaly tends to outbreed itself in families Once it has disappeared from a family it does not appear to recur The condition is presumed to be due to a defect in the parental germ plasm a faulty anlage in the

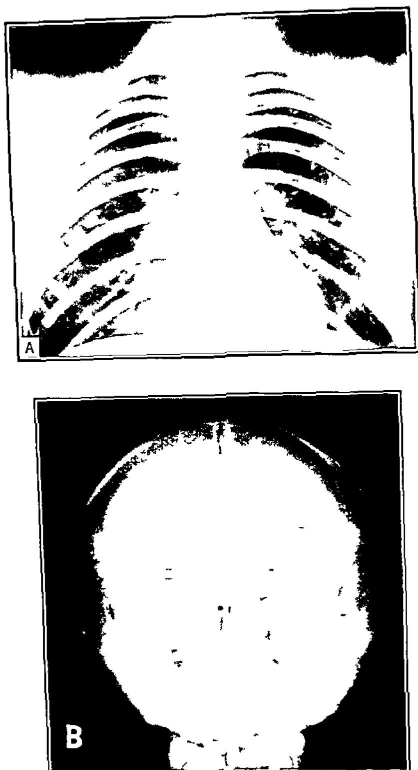


Fig 43 Cleidocranial Dysostosis A Chest The clavicles are absent and the scapulas are small and anomalous B Skull The head is large with increase of the transverse diameter The sutures are persistent and there are multiple wormian bones The face is small in relation to the head The patient is a male twenty seven years of age Despite the obvious anomalies he had always been in good health and had been able to perform all military duties during service in the armed forces

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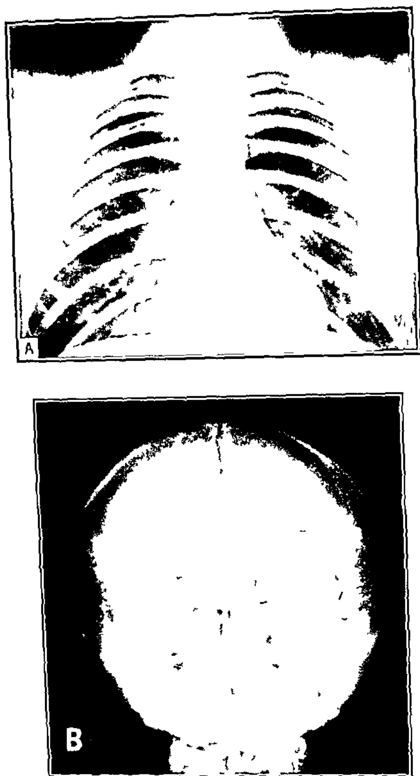


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skeletal system for membranous bone, particularly the clavicle and the cranium. Patients afflicted with this disease are usually small. The cranium is disproportionately large and there is prominence of the frontal and parietal bosses. The eyes are widely separated. The chest appears flattened in the area occupied by the outer portions of the clavicles. In many instances, the anomaly is first discovered during the course of a roentgen study for other conditions.

The bones of the skull may be divided into two large classes according to their method of ossification in cartilage or in membrane. In cleidocranial dysostosis, the membranous bones are principally involved. These comprise the frontal, parietal, squamous portion of the temporal bone, the upper portion of the occipital, and the petrous and mastoid portions of the temporal bone. The sphenoid (except the greater wings) and the eth-



FIG 44 Cleidocranial Dysostosis. The clavicles are small and underdeveloped with absence of the middle and outer portions. The remaining portions of the clavicle are indicated by the arrows.

moid are ossified in the cartilage of the base. The defective ossification of the bones of the vault produces the characteristic skull deformity. In infancy there are very large fontanels with wide sutures separating the bones. The fontanels may not close until very late in life. The changes are in very striking contrast to those in achondroplasia in which there are defects in the development of cartilaginous bones. Many of the defects seen in cleidocranial dysostosis are, however, present in rickets, *i.e.* delayed closure of the sutures and fontanels, prominent bosses and poor dentition. The head is very large and brachycephalic, the transverse diameter being increased with a cephalic index of over 80. Cephalic

$$\text{index} = \frac{\text{max. breadth} \times 100}{\text{max. length}}$$

The breadth is measured at the widest interparietal points, the length from the nasion to the posterior occipital point. The normal is under 80. The fontanels and widened sutures may persist throughout life. The bridge of the nose and zygoma are sunken

The sinuses are small or absent The palate is high and arched The neck is long, the shoulders narrow, the supraclavicular fossas indistinct, and the shoulders droop as in fractures of the clavicle Despite the clavicular defects, function is not greatly interfered with, those affected being unaware of anything wrong and being able to do strenuous work The range of motion of the shoulders is markedly increased Some individuals having these defects can bring their shoulders forward until they come almost in contact with each other under the chin The condition is not incompatible with a long and active life and cases have been reported in persons of all ages Our oldest patient was sixty six years of age and in good general health the youngest was twenty two years old

Roentgen Manifestations The roentgen findings are pathognomonic The skull shows generalized or diffuse areas of rarefaction, the frontal bones usually demonstrating the most ossification and the temporoparietal regions the least The sutures are broad, anomalous suture lines are present, and the fontanels are large and persistent The bones of the skull are large in proportion to those of the face The parietal and temporal bosses are markedly prominent with increase in the biparietal diameter of the head A depression may be present parasagitally between the bosses Semimembranous areas persist and a mosaic pattern with numerous Wormian bones may be noted The base is decreased in size, the squamous portions of the temporal bones lie diagonally, and the mastoids are directed more toward the midline than normally Hypertelorism is present, the eyes being widely separated

The clavicles may be partially or completely undeveloped on one side or bilaterally The defect in some instances is asymmetrical It may be mistaken for ununited fractures both clinically and roentgenographically, and important medicolegal complications may result The commonest defect is absence of the outer end of the clavicle, the sternal end being present The next most common finding is the presence of two separate fragments for each clavicle As a rule, the inner is the larger of the two and in some cases does not extend to the acromion The least frequent is absence of the sternal end with the acromial end being present Complete absence of the clavicles may occur but is rare The thorax is narrow superiorly and is best described as being cone shaped The ribs may slope downward to a greater degree than usual The sternum may be of peculiar shape and failure of the manubrium to ossify has been reported Many other skeletal defects have also been described The spine may show incomplete closure of the neural arches various fusions, and curvatures In some cases, there is stunting of the long bones and these bones are slender Rarely some or many of the tubular bones are overdeveloped, particularly the metatarsals and the second metacarpals The hands and feet show various abnormalities the most common and curious being the presence of epiphyses at both ends of the metacarpals and metatarsals particularly the second and fifth and an abnormally long second metacarpal The heads of the other metacarpals recede from the second in a regular step like fashion The epiphysis at the base of the second metacarpal is strikingly large There may be deficient and delayed ossification of the pubis in some instances this change being bilateral The inferior ramus of the pubis shares in the delayed and defective ossification There may also be coxa vara

The maxillas are hypoplastic but the mandible is of normal dimensions Fusion at the mandibular symphysis may be delayed or fail completely Abnormality of the teeth and jaws is practically a constant feature and is

characterized by delayed resorption of the roots of the primary teeth, failure of eruption of the permanent teeth and the presence of supernumerary dental structures The primary teeth show prolonged retention some of them for at least six years beyond the normal time for shedding The roots of the upper anterior and some of the posterior teeth undergo resorption While the primary teeth usually are lost by the time the patient reaches adult life most of the permanent teeth remain unerupted for long periods This is apparently due to the quality or character of the bone which fails to resorb in response to the eruptive force The roots of the permanent teeth may be short and slender

Pathogenesis A definite understanding of the genesis of the anomaly has not been established It is obvious from the number and variety of bones involved that the disease is much more generalized than was previously supposed Histologic studies have not been made in this condition, which accounts for the lack of definite knowledge as to its cause and nature The involvement extends to far more than merely the bones which develop from membrane The permanent teeth the facial bones the sternum, the scapulas the vertebrae the pelvic bones the long bones, the metacarpals, and the phalanges show defects which apparently involve both the ectodermal and mesodermal structures The causative agent appears to become operative at about the fifth week of fetal life and continues for an indefinite period thereafter, possibly extending into childhood The name cleidocranial dysostosis is inadequate and the term osteodental dysplasia is more appropriate being comparable with the related disorder of achondroplasia Jackson reports a study of the disease in which the affected members of the family over 20 years of age had lost all the teeth In a case recently reported by Scott and Banks the condition was noted *in a four year old* Negro girl In addition to the typical cleidocranial defects there was retardation of physical dental and epiphyseal growth as well as absence of the pubic bones At the age of four days there was only a shell of the frontal and occipital bones The parietal bones were membranous in type and could not be visualized Roentgen study of the lower extremities showed complete absence of the tibial and epiphyseal centers of the knee also of the os calcis the astragalus and the cuboid The patient was studied again at the age of four and one half years and the skull was found to be large with a wide transverse diameter and multiple Wormian bones The pelvis showed wide separation of the ilia and absence of the pubic bones as well as absence of the clavicles

Complications include pressure on the brachial plexus This can be relieved by removal of the outer fragment of the clavicle An accompanying calcification in the skin and soft tissues has been reported Syringomyelia has been mentioned as a rare complication The diagnosis can usually be made easily In the young patient with pseudarthrosis of a clavicle and thickening of the fragments without obvious deficiency in the skull roentgen examination of the pelvis usually makes the diagnosis clear

ADDITIONAL READING

Eisen D Cleidocranial Dysostosis Radiol *61* 21 1953

Jackson W P U Osteodental Dysplasia Cleidocranial Dysostosis Acta Med Scandinav *139* 292 1951

Soule A B Jr Mutational Dysostosis (Cleidocranial Dysostosis J Bone & Jt Surg *28* 81–102 1946

PUNCTATE EPIPHYSEAL DYSPLASIA, STIPPLED EPIPHYSES CHONDRODYSTROPHIA CALCIFICANS CONGENITA, CHONDRODYSTROPHIA PUNCTATA CHONDRODYSTROPHIA CALCAREA CONGENITA

The name chondrodystrophia calcificans congenita was first applied to the disease by Raap in 1942 It is discussed in the literature under the names of punctate fetal chondrodystrophy, dysplasia epiphysilias punctata, stippled epiphyses and chondroangiopathia calcarea seu punctata It has been termed calcinosis universalis but this is incorrect The condition belongs to a large, poorly defined group of bone and cartilaginous embryonic malformations which are classified as chondrodystrophies This group has been subdivided into many poorly defined secondary classifications which merge imperceptibly into one another However, it possesses many characteristics which do not occur in any of the other similar lesions and therefore should be classified in its own special group The first roentgen observations were described in 1914 by Conradi whose description is classical None of the subsequent discussions have added materially to his observations The disease is usually discovered soon after birth because of obvious deformities, although in some instances it is first noted during the early months or years of life due to lowered general health and decreased growth of the patient Careful study of the fetus *in utero* during the later months of pregnancy may permit of the establishment of the diagnosis before birth

Roentgen Manifestations The diagnosis is made only by roentgen studies The condition is characterized by the presence of multiple finely stippled, diffuse dense calcifications in the ossification centers of the bones of the tarsus and carpus and in the regions of the epiphyses in any portion of the skeleton The calcific depositions are discrete and sharp and may measure a few millimeters in diameter or be very extensive in size There is in many instances an appearance of semicalcified, amorphous density in the regions adjacent to the calcific shadows The calcifications may occur in any portion of the skeleton in which cartilage is normally present While the calcific depositions are widely distributed throughout the body in most cases, there have been a few instances recorded in which only the lower extremity or the tarsal areas were affected The calcifications in many cases appear at a much earlier age than that at which ossification normally is expected in the affected epiphyses and ossification centers Many of the patients die in infancy or early childhood The cause of death is usually an infection such as pneumonia tuberculosis or pyelitis Hence the progress of the disease can seldom be followed for long periods of time In the few individuals who survive for longer periods, the calcified material appears to coalesce and decrease in extent and tends to disappear according to Raap at the age of three or four years

While the majority of cases show extensive involvement of the skeleton, the most frequent and severe calcifications occur in the tarsal and carpal areas All of the bones preformed in cartilage may be involved The calcific depositions occur at the ends of the long and short tubular bones, the large flat bones of the pelvis the chondral ends of the ribs and the vertebral ossification centers The calcified stippling may appear in cartilaginous areas which are never ossified at birth and have been reported in the costal cartilages and the intervertebral discs The patella presents

characterized by delayed resorption of the roots of the primary teeth, failure of eruption of the permanent teeth and the presence of supernumerary dental structures The primary teeth show prolonged retention some of them for at least six years beyond the normal time for shedding The roots of the upper anterior and some of the posterior teeth undergo resorption While the primary teeth usually are lost by the time the patient reaches adult life most of the permanent teeth remain unerupted for long periods This is apparently due to the quality or character of the bone which fails to resorb in response to the eruptive force The roots of the permanent teeth may be short and slender

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is broadening of the shafts with a tendency to flaring of the metaphyses In many instances, there is uneven longitudinal growth of the radius and ulna and of the tibia and fibula relative to each other This results in lateral or medial deviation of the wrists and the feet There may be shortening of the extremities due to decrease in length of the femurs and humeruses, the peripheral bones not being shortened Dwarfism occurs only in the severe cases As the patient grows older, there is usually progressive ossification of the epiphyseal cartilage The calcific stipplings merge into each other and gradually decrease in number The epiphyses may be deformed and present an appearance similar to that in epiphyseal necrosis The metaphyses may appear mottled and irregularly rarefied with thinning of the cortex The picture in later life is easily confused with residuals from osteochondritis

Clinical Manifestations Stiffness of the joints is an important manifestation The patients maintain the main joints such as the hips, knees, ankles, shoulders, elbows and wrists in semiflexion and there is a moderate but definite resistance to extension This is best explained by Fairbank's report of a case seen at autopsy in which the muscles near the joints were largely replaced by fibrous tissue There is no definite familial tendency although Raap described 4 cases in siblings, two of which were in twins Maitland also discovered the disease in 2 sisters whose father had a congenital absence of several phalanges in the hands and toes In the cases reported by Tisdall the parents were first cousins In none of the other reported cases has there been any suggestion of hereditary or familial tendencies in the parents or the siblings Another anomaly which may occur in some cases is a thin, flabby, and scaly skin There is a tendency to a peculiar formation of the skull with oxycephaly, bossing of the frontal bones, and sinking and flattening of the bridge of the nose These occur relatively frequently but not regularly There is no sex preference In 17 of the reported cases in which sex was mentioned specifically 8 were in boys and 9 were in girls The children gain weight slowly and are retarded Most of the patients have died during the first year of life

Pathology Histologic studies are rare Fairbank mentioned areas of mucoid degeneration and the formation of cystic spaces in the cartilaginous epiphyses, especially near the articular surfaces There may be invasion of the degenerated areas by blood vessels and a core of fibrous tissue The vertebral ossification centers show a lack of the usual orientation of cartilage cells and of normal calcification and ossification The principal error appears to be of the same order as in achondroplasia There are circumscribed, polymorphous deposits of chalk in the cartilage with larger confluent areas of calcification in the adjacent tissues In some portions new bone formation replaces the chalky areas The zone of ossification between the bone and cartilage at the epiphyseal line appears diminished The flecks of calcification may appear stellate in shape Fibrous degeneration occurs in the muscles near the joints

Raap describes a case first seen by him in 1938 The roentgenograms were those of a male child age ten months The long bones showed flaring of the distal ends of the ulnas and tibias together with a mottled, granular appearance in the ankles wrists and other joints The change in contour followed that of the bony structures in normal ossification but presented angular rather than rounded densities Roentgenograms of a twin who had died were made and the joints showed similar changes Microscopic study of the bones of the ankle showed an increase of fibrous tissue of

an irregular mottled appearance The thyroid cartilage may show dense calcification In the region of the ends of the long bones, the calcifications extend beyond the areas which one would normally expect to be occupied by cartilages and in cases seen at autopsy there are indications that the muscular and supporting tissues near the joints are also affected The centers of ossification appear grossly deformed It is believed that the calcifications at the ends of the long bones probably represent flattened and widened centers of ossification In some instances the calcific depositions fuse to form a more nearly normal appearing single center of ossification In addition to the calcifications there is also definite evidence of chondrodystrophy of the long bones The long bones are shortened and there

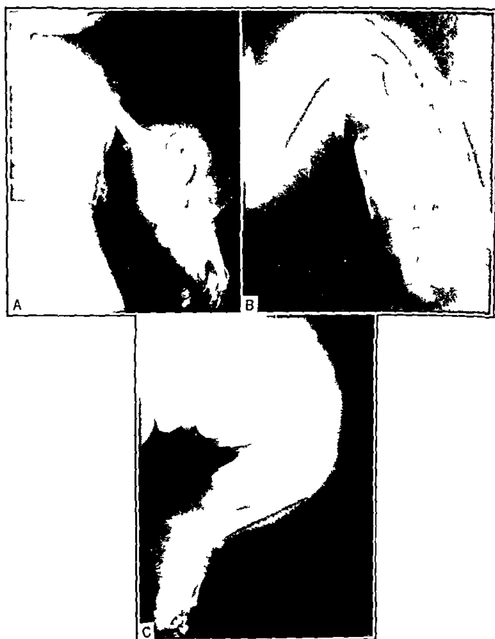


FIG 45 *Stippled Epiphyses* *A The right arm* *B The left arm* *C The left leg* There are multiple finely stippled dense calcifications in the regions of the epiphyses and the ossification centers of the carpus and tarsu The long bones are shortened and the shafts are widened There is maldevelopment of the bones of the right forearm There are multiple calcific nodules in the chest most probably in the costochondral regions

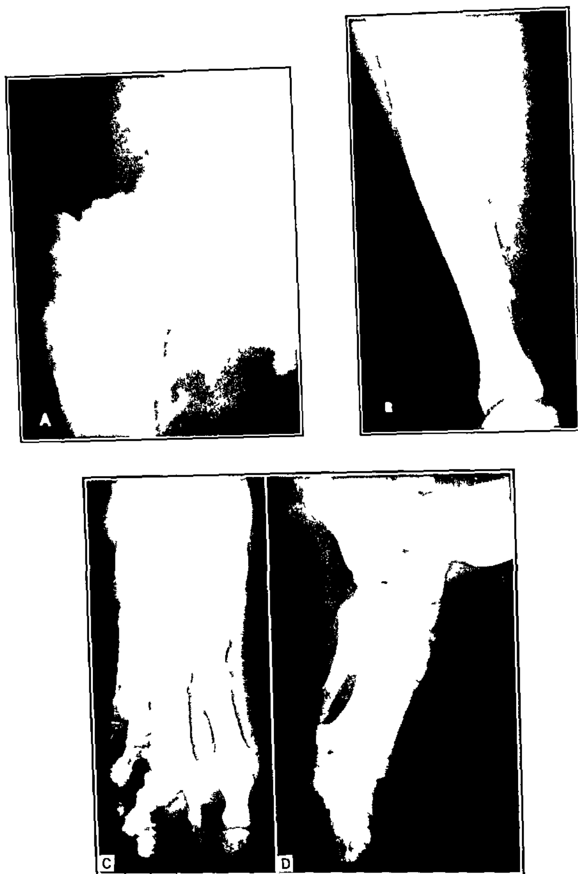


Fig 46 Melorheostosis A The hip B Lower leg C Foot anteroposterior view D The foot oblique view There are multiple large areas of calcification in the soft tissues of the hip leg and foot The calcific shadows are irregular in outline and have the appearance of molten wax the characteristic finding in melorheostosis There is cortical thickening involving the long bones especially the tibia

periosteal character Directly under this periosteum, a formation of cartilage was present The deeper cells were acquiring a cartilaginous character The deepest layers of cells formed osteoid tissue which was partially calcified According to Cocchi there is a disturbance in the vascularization of the epiphyseal cartilage and the cartilaginous anlage of the small bones of the extremities The cartilaginous columns show varying degrees of divergence from their usual parallel arrangement Whether the primary anomaly is within the blood vessels or the vessels merely parallel the abnormally coursing cartilage columns is not known The epiphyseal cartilage and the anlage of the small flat bones show marked vascularization and irregular areas of mucous degeneration

Differential Diagnosis The differential diagnosis of stippled epiphyses is not difficult The roentgen demonstration of multiple calcific densities throughout the cartilaginous system in a newborn infant is pathognomonic of the disease Stippling of the epiphyses may occur in association with hypothyroidism, the condition being known as cretinoid epiphyseal dysgenesis However this condition occurs later in life usually from the second year onward, has a less universal distribution, and the individual foci of calcification tend to be larger As the child with hypothyroidism grows older the epiphyses are well formed although their development may be delayed Multiple foci of ossification within an epiphysis occur as a normal variant Stippled epiphyses differs from chondrodystrophy in that thickening of the shafts of the long bones is not present and there is no shortening or broadening of the diaphyseal ends of the long bones

ADDITIONAL READING

MAITLAND D G Punctate Epiphyseal Dysplasia Occurring in Two Members of the Same Family Brit J Radiol *12* 91 1939

MASEKILDE F Stippled Epiphyses Acta Radiol *37* 291 1952

RAAP G Chondrodystrophia Calcificans Congenita Am J Roent *49* 77 1943

MELORHEOSTOSIS

In 1922 Leri and Joanny described a previously unreported anomaly characterized by an unusual distribution of hyperostosis along the side of the bones of an extremity producing a picture which resembled the tallow drippings down the side of a candle To this flowing hyperostosis of a limb they gave the name melorheostosis The term osteitis eburnans monomelica has been suggested as it is indicative of hardening of the bone involving a single limb Another name which has been used is osteopathia hyperostotica condensans The condition is sufficiently rare so that even today only solitary cases are recorded It differs from many other bony anomalies of an asymptomatic nature which are discovered by accident in that it constitutes a specific syndrome

Clinical Manifestations The symptomatology varies with the location extent, and activity of the disease The process is purely local and is not associated with generalized constitutional manifestations Not infrequently the condition has been discovered by accident before the onset of symptoms However pain is a common complaint and is present in practically every case It is rarely severe and is not constant Characteristically the pain is vague boring and disappears on rest When the condition begins early in life there is abnormal curvature of the involved

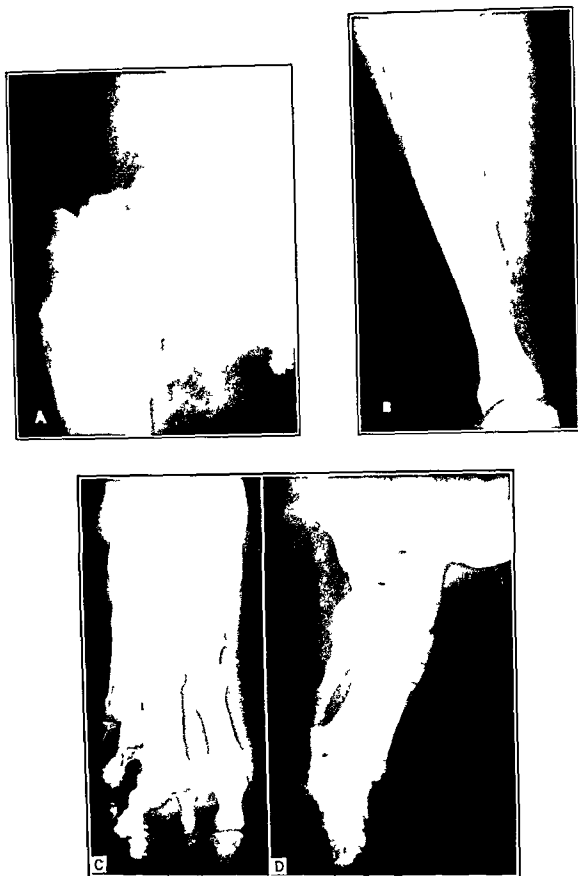


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pelvis, one side of both lower extremities, and all four extremities However, the distribution is not entirely without pattern inasmuch as no cross arrangement has been noted In the early stages there are no characteristic manifestations In the case which has progressed sufficiently, one finds massive thickening of the bones of the extremities usually throughout its entire extent, increasing proximally to distally, and more marked as a rule on one side Similar bone thickenings may be present in the shoulder and the pelvis The bone accretion gives rise to secondary changes, humping and bowing of the shaft, shortening or occasional elongation, and striping within the cortex of the bone Particularly in the pelvis the striped pattern of increased bone density is arranged in parallel bands which radiate in the direction of the hip joint Sometimes spurs project into the soft tissues from the border of the acetabulum

Pathology There have been found cortical hyperostoses of dense sclerotic bone endosteally, periosteally, or both The condition appears to be a benign, progressive one There is atrophy of the involved bone Expansile enlargement has not been described On microscopic examination there is marked density of the bony lamellæ with overcrowding and bizarre formation There is usually a reduction in the number of osteoblasts The Haversian system is distorted and bizarre and in many instances is compacted, although occasionally it is rarefied There is no evidence of inflammatory changes Fibrotic infiltration of the trabecular structure of the bone and fibrotic replacement of the marrow is considered to be the fundamental or primary pathology There is a wide divergence in the proportion of osteoblasts to osteoclasts This indicates that the condition has alternating osteoblastic and osteoclastic phases The histopathology is dependent upon the degree of activity, the stage of the disease and the portion of the bone from which the biopsy is taken The picture on microscopic examination is similar to that of several other hyperostotic bone lesions This suggests that these conditions may have similar basic or underlying pathologic changes although the etiology may be different

Hoffken and Hein have published the first autopsy report in melorheostosis The authors had the opportunity of obtaining a biopsy a year prior to the patient's death The findings at this time and at the autopsy were compared and there were several distinct differences In the specimen taken at biopsy the bone was poor in calcium while at postmortem it was extremely rich in calcium The osteoblasts were arranged in several layers in the biopsy specimen whereas after death one year later they were markedly diminished, formed only one layer and the osteoclasts had disappeared It appeared that the sclerosing process of the bones which was very active a year previously had now been completed This is believed to explain the contradictions in the literature in regard to the microscopic features of the condition as described on previous studies of biopsy specimens In some cases there had been noted the presence of osteoblasts Therefore it is reasonable to conclude on the basis of the findings in this case that these are different stages of the same process Putti considered the disease to be the result of local vascular obliteration produced by neurosympathetic vasomotor stimulation due to the fact that increased density occurs in the bony architecture after vascular curtailment It is known that an increase in the blood supply to a bone results in decalcification while a decrease in the blood supply produces an increase in density of the bone or hyperostosis The periosteum does not contain a specific osteogenic function and the osteoblast is not endowed with special

bone Shortening of the limb may be present When the process extends beyond the confines of the periosteum, stony hard swellings and deformities may become palpable and visible The affected limb may feel heavier and appear larger than its fellow despite the fact that there is frequently concomitant muscle atrophy Pressure on the nerves and the blood vessels produces paresthesia neuralgia, venous congestion and edema The symptoms are sporadic and may disappear for months or years The involved parts undergo atrophy Motion may be restricted by mechanical obstruction and in some instances there is immobilization without actual ankylosis due to thickening of the contiguous articular surfaces or heterotopic ossification in the para articular structures Originally, it was believed that the condition was confined to a single extremity While this is true as a rule cases have been reported in which both limbs and other portions of the skeleton have been involved An important characteristic is the selectivity or predilection for one side of the bone such as the femur or the humerus or to a single bone where two bones lie parallel to each other as in the case of the tibia and fibula or radius and ulna In those instances in which the process extends from the carpus or tarsus to the phalanges a linear segment only is affected The extent of involvement is dependent upon the stage at which the case was first observed and the rapidity with which the process had advanced In some instances the disease remains localized to a single digit or small segment of the lower end of one long bone whereas in others there is involvement of all four limbs part of the pelvis the lumbar spine and the skull Diagnosis can be made only on the basis of the roentgen findings

Roentgen Manifestations The first manifestation is that of irregular thickening of the inner border of the cortex of one side of the bone The roentgen examination may reveal a dense, hypertrophic irregular linear or plaque like mass in the region of the marrow In the early stages only the inner border of the hyperostotic mass is irregular As the disease progresses, there is distortion and thickening of the periosteum The joints in the region involved remain unaffected even in cases in which the process involves both sides of the joint Many cases have been reported with heterotopic deposits of dense calcareous strips in the adjacent soft tissues particularly in the region of the joints In the advanced stages there is replacement of the normal bone reticulation by a dense stony opacity with a peculiar patchy linear distribution and selective localization to a single side of a bone or to one of paired bones The areas of hyperostosis are dense and structureless The deposits may be quite small or may extend over an entire long bone or limb giving the appearance of guttering wax of a candle The new bone is usually deposited cortically in the long bones and endosteally in the short bones The distribution is peculiarly segmented in pattern The segments do not correspond with the course of the vessels or the nerves Soft tissue deposits of the bony material frequently occur but the joint margins are not affected The soft tissue depositions form dense irregular plaques in the regions adjacent to the margins of the bones

In a case described by Murray there were typical widespread changes involving the left limb with dense patches elsewhere on the left side of the body particularly the sacro iliac joints the bodies of the fourth cervical and ninth dorsal vertebræ the 8th and 9th ribs and the superior and inferior angles of the scapula There have been instances reported with involvement of the upper and lower extremities including one half of the

factor with a selective dystrophy of the mesodermal tissues There has also been noted in certain instances a hormonal disturbance, particularly hyperfunction of the eosinophilic cells of the pituitary The onset of the syndrome appears to be *in utero*, hence it is unlikely that endocrines could affect the fetus because of compensation by the mother The theory of a congenital muscular dystrophy has been discredited The muscular disturbance in arachnodactyly *is not* progressive muscular atrophy It has been considered by some authors that there is a defect in the germ plasma The hereditary factor is stressed by practically all observers In a large percentage of the cases there has been a hereditary background The symptoms and signs are referable to various systems of the body

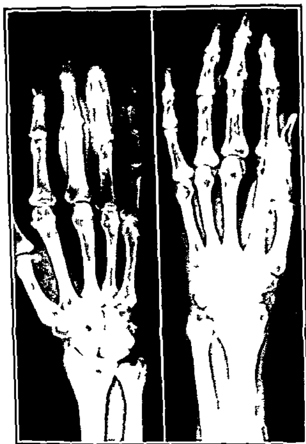


FIG 47 Arachnodactyly The bones of the hand are markedly elongated and show cortical thinning with slight generalized osteoporosis

The bony structures are markedly affected, the fingers being long and tapering The metacarpals and metatarsals are most extensively involved There is a severe reduction in the amounts of subcutaneous fat, imparting the appearance of malnutrition The patients are usually double jointed due to relaxation of the ligaments The ears are prominent with pointed tips The palate is high and arched Kyphosis and scoliosis with sternal deformities are frequently present Webbing of the fingers is common The facial appearance is peculiar A long drooping jaw, eyeglasses, thin face and long ears give the patient a mournful expression There is poor development of the secondary sexual characteristics The basal metabolic rate may be decreased Pneumonia is the usual cause of death and is probably due to the chest and spine deformities in association with the anomalies of the lungs Mental retardation is rare The eye find

power to lay down new bone The deposition of lime salts is a physical phenomenon under the influence of the endocrines and the local control of the enzymes, particularly phosphatase on the chemical balance of tissue fluids Diminished blood supply resulting from vascoconstriction thrombosis embolism, low metabolism, or partial occlusion by twisting, kinking or pressure leads to local asphyxia with attendant fibrosis Calcification takes place in fibrous tissue of impaired vascularity as in myositis ossificans, chronic osteomyelitis and syphilitic osteitis

Differential Diagnosis The condition has been confused with other hyperostotic bone lesions such as syphilis osteopetrosis (Albers Schonberg disease), osteopoikilosis osteitis deformans osteitis fibrosa disseminata, polyostotic fibrous dysplasia, myositis ossificans osteogenic sarcoma carcinomatosis chronic osteomyelitis traumatic or infectious ossifying periostitis and the non suppurative ossifying periostitis of Garre

Prognosis and Therapy The disease has a slow and insidious onset, progresses slowly with periods of temporary or permanent arrest although never showing reversal to normal bone and tends to be chronic Malignant degeneration does not occur Pathologic fractures have not been reported This is most probably due to the characteristic partial involvement along one side of the bone or one of a pair of bones and also the fact that the sclerotic replacement of the Haversian system is on a lamellar pattern which results in retention of the resilience of the bone The pain disappears on rest and physiotherapy In long standing cases there is limitation of motion of the joint In many instances operative removal of mechanical causes of obstruction may become necessary

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ARACHNODACTYLY MARFAN S DISEASE

This syndrome was first reported in the literature by Marfan in 1896 who termed the condition spider feet The name hyperchondroplasia has also been suggested Because of the spider like appearance of the digits when extended it has been termed arachnodactyly There have been about 200 cases recorded in the literature The first reference in the American literature was in 1926 by Piper and Irvine Jones They found the lesion in association with congenital heart disease The condition is doubtless much more common than is indicated by the relatively small number of cases which have been reported In those which have come to autopsy various cardiac lesions and other anomalies have been found including a patent foramen ovale and a two lobed right lung an enlarged heart due to a patent foramen ovale an interauricular septal defect with failure of division of the lobes of the lungs fenestration of the mitral valve and a small frontal lobe of the brain There also have been described two cases with aneurysmal dilatation of the ascending aorta proximal to the aortic arch one of which had a dissecting aneurysm

There are numerous theories as to the cause of the disease the chief of which are that it is (1) a constitutional aberration of the anlage (2) a neurological disorder similar to syringomyelia and (3) due to a hereditary

corded as occurring in most cases According to Miller, the typical deformity involves the lower dorsal region and comprises a kyphoscoliosis in which the kyphotic element predominates The scoliosis is considered due to multiple tumors of the spinal nerve roots but may also be secondary to coexisting vertebral anomalies or disorders of growth of an extremity

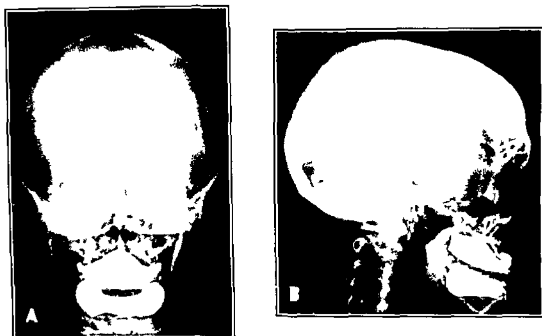


Fig 48 Neurofibromatosis *A* Anteroposterior projection The sphenoidal ridges and the zygomaticofacial portions of the sphenoidal bones are completely absent The lateral margins of the orbits appear thinned *B* Lateral projection The shadows of the sphenoidal ridges the anterior clinoids the anterior portion of the sella and the greater and lesser wings of the sphenoid are absent (Courtesy of John F Holt, M D)

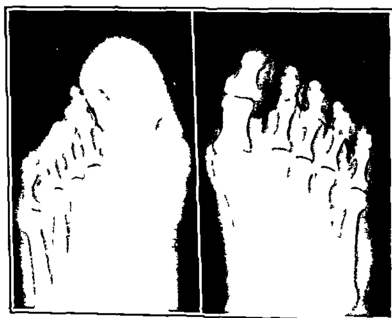


Fig 49 Neurofibromatosis The soft tissues of the great toe are markedly increased in thickness and density Within this density there are numerous small rounded nodular shadows which represent the neurofibromata The phalanges of the great toe show irregularity of outline and erosions The contralateral foot which is not affected by the disease is shown for comparison

ings are of significance and are important in diagnosis They are deeply set and myopic About half the cases have a bilateral dislocation of the lens The pupils are maintained in mid constriction and dilate poorly with atropine Great stress must be placed on the relatively high incidence of heart disease abnormalities of the heart and lungs being present in about half of the cases

NEUROFIBROMATOSIS

Von Recklinghausen presented his classical description of neurofibromatosis in 1882 The condition is very variable and is not infrequently associated with bone changes hence roentgen methods of diagnosis are extremely valuable in diagnosis It appears unusual to find mesenchymal defects in a disease which is believed by most authorities to be neuroectodermal in origin The paradox is best explained by the theory of Yakovlev and Guthrie They discuss a group of related congenital malformations which affect the ectodermal structures electively Three syndromes are included in this group under the heading of neurocutaneous syndromes (1) Recklinghausen s neurofibromatosis (2) tuberous sclerosis, and (3) cephalotrigeminal angiomatosis the Sturge Weber syndrome The fundamental fault in these conditions is considered a congenital ectodermal dysplasia originating early in embryonic life when the cells of the body have polyvalent possibilities of differentiation There is an associated secondary phenomenon comprising hyperplasia and actual new growth of mesodermal tissue and hyperplasia of the blood vessels, in some instances so pronounced as to constitute a hemangioma

In the well established case of neurofibromatosis with multiple cutaneous or subcutaneous tumors and numerous large *cafe au lait* spots diagnosis is easy However, it is not always possible to establish a definite diagnosis as one or more pigmented nevi may occur and closely resemble the *cafe au lait* spots of neurofibromatosis Many observers are of the opinion that the tumors of neurofibromatosis are specific pathologic entities and differ from the usual solitary neurofibroma termed the perineural fibroblastoma In order to establish the diagnosis of neurofibromatosis it is essential to have a combination of typical cutaneous and subcutaneous lesions with or without bone involvement

The characteristic bone defects in neurofibromatosis have been studied exhaustively by Holt and comprise the following (1) erosive defects due to the presence of neurofibromas contiguous to the bone (2) scoliosis (3) disorders of growth comprising overdevelopment underdevelopment and trophic changes, (4) bowing and pseudarthrosis of the lower leg (5) intraosseous cystic lesions and (6) miscellaneous anomalies

(1) *Erosive Defects* These are usually saucer shaped areas of bony erosion due to the presence of neurofibromas in the tissues adjacent to the bone They are most common in the spine and occasionally occur in the extremities the clavicle the scapula and other bones In some instances there are symmetrical erosive defects of the inferior aspects of the ribs which resemble the pathognomonic sign of coarctation of the aorta and are due to bead like distribution of multiple small neurofibromas of the intercostal nerves

(2) *Scoliosis* Abnormal curvature of the spine is the most common osseous abnormality in neurofibromatosis and by many observers is re

examination of the skull shows absence not only of an orbital wall but in some instances of practically the entire sphenoid bone Despite the extensive roentgen manifestations there is no clinical evidence of an intracranial tumor There is no logical explanation for these manifestations Spina bifida, club foot, congenital dislocation of the hip and other anomalies have been recorded in cases with neurofibromatosis

Involvement of the bones of the skull is common The lesions are protean and comprise bulging of circumscribed areas, peculiar vascular designs, enlargements of the sella, the orbit and the orbital canal, gross asymmetries of various types, subperiosteal cysts, bone atrophy, and



FIG 50 Neurofibromatosis There are multiple neurofibromata in the soft tissues of the upper arm and chest There is erosion and absorption of the outer end of the clavicle the spine of the scapula and the acromion process The soft tissue masses in the chest wall may be mistaken easily for pulmonary tumors



FIG 51 Neurofibromatosis of the Shoulder There is diffusely increased density of the soft tissues in the region of the shoulder and the axilla There are deformities and erosions of the scapula and the outer end of the clavicle and separation of the acromioclavicular joint

(3) *Disorders of Growth* The most commonly encountered anomaly is overgrowth of one or more bones There is usually extensive soft tissue swelling and pigmentation of the skin overlying the involved bone On histologic examination there are plexiform or cirsoid neurofibromatous tissues infiltrating the soft tissues This produces a bizarre topical malformation of an elephantiasic variety The periosteum may be invaded diffusely by the neurofibromas producing a characteristic type of wavy periosteal new bone formation The overgrowth of bone in neurofibromatosis may be both longitudinal and circumferential and in children is frequently accompanied by accelerated maturation of the epiphyseal ossification centers While the mechanism of these changes is not definitely understood, it is most probably due to local hyperemia resulting from stasis of lymph There also may be definite undergrowth of the tubular bones of the extremity with moderately extensive soft tissue neurofibromatous infiltration Shortening of the growing bone is due to invasion by the neurofibromatous tissue of the plate of the epiphyses Skeletal trophic changes occur in the infiltrating plexiform neurofibromas There is narrowing sinuousity and progressive decrease in size of the long or short tubular bones ribs clavicles and ilums In some cases the trophic phenomena are weird and cannot be explained Holt describes a withered fibular stump and narrowing or tortuosity of the clavicle in a patient with neurofibromatosis

(4) *Congenital Bowing and/or Pseudarthrosis of the Tibia* Congenital anterolateral bowing of the tibia is not uncommon in children with neurofibromatosis In consequence all patients with congenital deformities of the tibia should be observed over a period of years because of the probability of soft tissue tumors developing later Pseudarthrosis of a kyphotic tibia and fibula may result after a minor injury Similar changes have occurred in the forearm following trauma True neurofibromatous tissue is seldom found at the site of a pseudarthrosis following fracture of a congenitally bowed tibia It is believed that the original fault in congenital bowing occurs in the early pre ossification developmental phase of the limbs Pseudarthrosis does not occur in every case

(5) *Intraosseous Cystic Lesions* There are many cases on record of histologically proven primary intraosseous neurofibromatosis The neurofibroma may arise from a periosteal nerve and erode the bone Simultaneously there is stimulation of the osteogenic layer of the periosteum with formation of a shell of bone over the neurofibroma In consequence the tumor lies within the cortical or cancellous bone and appears to have arisen from this portion of the bone where is actually it is periosteal in origin Localized cortical defects in the growing long bones of many children should not be mistaken for intraosseous neurofibromas Not infrequently there are widespread intraosseous cyst like lesions of varying size The lesions are most apt to be found within the metaphyses of the tubular bones The cystic bone lesions may be associated with ill defined foci of calcification in the soft tissues due to necrotic centers of subcutaneous neurofibromas In some instances the cystic lesions regress partially and spontaneous disappearance may occur These manifestations emphasize the fact that neurofibromatosis is a congenital dysplasia rather than a true neoplasm

(6) *Miscellaneous Associated Anomalies* There have been reported absence of a portion of one orbit associated with pulsating exophthalmos Approximately 35 cases of this type have been recorded The roentgen

examination of the skull shows absence not only of an orbital wall but in some instances of practically the entire sphenoid bone Despite the extensive roentgen manifestations there is no clinical evidence of an intracranial tumor There is no logical explanation for these manifestations Spina bifida, club foot, congenital dislocation of the hip and other anomalies have been recorded in cases with neurofibromatosis

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erosions The bony defects may be due to localized destruction of bone by neurofibromata or are associated with congenital anomalies This is particularly true in neurofibromatosis with defects of the orbital wall The defect in the orbital wall may permit of free communication between the intracranial cavity and the orbit Temporal and parietal bossing is common in neurofibromatosis The bulging may be unilateral and on the side of the tumor Roentgen evidences of increased intracranial pressure are frequent in children Other types of tumor both of the brain and the body in general, are frequent in association with neurofibromatosis Gliomas and meningiomas are often associated with neurofibroma Exophthalmos is a frequent manifestation and may be due to the presence of retrobulbar neurofibroma, an associated tumor of other type such as a glioma of the optic nerve or encroachment on the orbital structures by the intracranial contents In a case reported by Tucker and Carpenter there was a localized neurofibroma with associated marked overgrowth of the affected limb within a period of one year Melorheostosis has been described in a small number of cases There is no definitely proven relationship between this anomaly and neurofibromatosis It is known that melorheostosis is a bizarre typically unilateral skeletal hypertrophy or flowing hyperostosis

Joint changes are infrequent hence have received relatively little attention in the literature The articular manifestations comprise three principal groups (*a*) dysplasia (*b*) sclerosis of the articular portion of the bones and (*c*) secondary osteoarthritis (*a*) Dysplasias The dysplasias may occur at the hip and comprise coxa valga with a shallow acetabulum coxa vara, and congenital dislocation In the foot and ankle there may be anomalies of the calcaneus associated with distortion underdevelopment and displacement of the tarsal bones congenital rocker foot flat foot due to misshapen tarsal bones and lateral displacement of the talus and calcaneus In the hand there may be hypertrophy of the bones of the first and second digital rays associated with expanded bone ends (*b*) Sclerosis of the articulating aspect of the bone This may accompany the dysplasia or may be present as the sole abnormality in various joints In the hip there is sclerosis of the acetabulum associated with multiple radiolucent areas In the ankle there may be sclerosis of the lower segments of the tibia and the talus There may occur sclerosis of the distal end of the radius and sclerosis and hypertrophy of the carpal bones (*c*) Secondary osteoarthritis may occur in the presence of coxa valga or normally developed joints subjected to abnormal forces There have been no recorded cases of lesions of the shoulder or elbow with neurofibromatosis

Etiology There is no important difference in incidence in the sexes and there is no significant racial distribution The age of the patients varies from birth to 77 years In many instances there is a history of an exacerbation or first appearance of the tumors at about the time of puberty There is an apparent relationship to pregnancy and a high familial incidence Von Recklinghausen s disease is now known to be a congenital anomaly in which the cutaneous tumors and pigmentation are only two manifestations of a condition which may involve any or all systems of the body The hereditary nature has been conclusively established The controlling factor is a dominant one which follows regular mendelian principles The underlying cause of the disease is believed to be a defective germ plasm Masson believes the cellular origin of the condition is in the sheath of Schwann cells and proposes the term schwannoma Penfield and Cowe believe the tumor is produced from fibroblasts and preserves the characteristics of connective tissue

Pathology Neurofibromatosis is a disease which involves the sheaths and enveloping membranes of the nervous system The lesion is characterized in the nerve sheath tumors by typical palisading of the cells and streaming of cell bundles The tumors have been named neurilemoma nilemoma, neurinoma schwannoma and perineural fibroblastoma It has not been determined definitely whether these tumors arise from the mesodermal elements of the sheaths of the nerves or are derived from cells of neuroectodermal origin of the sheath of Schwann

SKIN MANIFESTATIONS There are pigmented nevi and these appear to be true nerve ending neuromas which develop from the peripheral sensory apparatus The *cafe au lait* spots which occur in neurofibromatosis and ordinary pigmented nevus cannot be differentiated by histologic methods In some instances there are also soft pedunculated tumors which appear to arise from the skin and are termed molluscum fibrosum These are not uncommon in neurofibromatosis and are of interest to the radiologist as they may cast soft tissue shadows on the roentgenogram

LESIONS OF THE PERIPHERAL NERVES The tumors of the peripheral nerves may be discrete neurofibromas Nerve fibers enter and traverse the tumors In some instances the peripheral nerve lesions are profuse Hypertrophy of the bones and periosteal new bone formation may occur in association with this type of lesion of the nerve

INVOLVEMENT OF THE CENTRAL NERVOUS SYSTEM In a high percentage of cases variously estimated as about 10 per cent there are associated intracranial tumors These may include meningiomas astrocytomas, spongioblastomas acoustic neuromas and undifferentiated gliomas In several instances the intracranial tumors may be multiple Tumors of the spinal cord and the spinal nerve roots also are very common These tumors present a dumb bell shape projecting to either side of the spinal nerve canal root and produce erosion of the pedicles and widening of the intervertebral foramens

MALIGNANCY Malignant degeneration in a neurofibroma is not infrequent and has been estimated as from 7 to 22 per cent The lesions are found to be neurofibrosarcoma fibrosarcoma or spindle cell sarcoma

BONE INVOLVEMENT Bone changes usually occur in combination with characteristic pedunculated soft tissue tumors and coffee colored blotches in the skin The presence of nerve tissue in compact and cancellous bone has been amply demonstrated in man Sympathetic and cerebrospinal nerves are present in the periosteum and constitute the chief nerve supply to the bones Small trunks have been described in the region of the nutrient vessels From the periosteal nerves minute fibers enter the underlying bones and extend to the Volkmann s canals Nerve endings have been found in extra osseous connective tissues but have never been identified in similar tissue within the bone marrow In connective tissue, the nerve fiber loses its myelin and is represented only by Schwann's nuclei It appears that the nerves in the bones terminate in the same way There are also nervous elements in the marrow

In one instance described by Weber surrounding and completely covering the tibial shaft was a diffuse mass of thickened neurofibromatous periosteum Areas of cranial hyperostosis have been observed in relation to overlying neurofibromatous soft tissue In the absence of neuromas overlying the sclerotic bone which might have produced these changes by chronic irritation it may be supposed that the effect is due to the neurofibromatous tissue in the marrow cavity The relationship of neurofibro

mas to peripheral nerves has long been recognized and the bone changes associated with the development of neurofibroma from the nerves of the periosteum are clearly understood.

Differential Diagnosis. The bone manifestations of neurofibromatosis are very variable. In consequence neurofibromatosis should be considered in the differential diagnosis of all bizarre skeletal diseases before a definite diagnosis is established. There should be other evidence of von Recklinghausen's disease and the bone lesions should fit into the entire picture. The presence of the neurofibromata adjacent to the vertebra with absorption of the vertebra produces very severe deformities. The lesions in the cortex of the bone may be merely scalloped defects and in many instances these appear and disappear without evidence that they are associated with neurofibromata. Fibrous dysplasia may produce similar defects in the bones and must be considered in differential diagnosis in all doubtful cases. The clinical diagnosis of neurofibromatosis is usually obvious because of the presence of the characteristic *cafe au lait* spots and the fibromata. The neurological signs vary with the particular nerves involved. Skeletal changes are of two types. More than one half of the patients have scoliosis and 7 per cent show bone changes which may be lytic or proliferative. The erosive changes may occur with other tumors, aneurysms and coarctation of the aorta. Overgrowth of individual bones occurs in hemangioma, lymphangioma, chronic osteomyelitis, hemihypertrophy and occasionally in hemophilia as a result of recurrent hemarthosis. Chronic osteomyelitis and syphilis may produce similar changes in the periosteum. Atrophy and underdevelopment of bone occur in postparalytic states and debilitating diseases. It is important to note that there is no osteoporosis in the atrophy associated with neurofibromatosis. In osteogenesis imperfecta the changes are more generalized. The cystic lesions which occur within the bones may resemble fibrous dysplasia, bone cyst, hyperparathyroidism, Ollier's disease and reticulosis.

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TUBEROUS SCLEROSIS

Tuberous sclerosis is an heredofamilial disorder of which the distinctive pathologic feature is the development of multiple glial nodules within the brain, especially over the gyral summits and in the ventricular linings. The nodules may become the seat of calcific deposits of sufficient density to be demonstrable by roentgen methods. They manifest themselves clinically by the onset of mental deficiency and epileptic seizures in infancy or early childhood. To these cerebral lesions are usually conjoined one or more of various extracerebral anomalies and neoplasms of which a facial adenoma sebaceum is so frequent as to be almost indispensible for diagnosis of the disorder. This reddish to reddish brown nodular eruption has a characteristic "butterfly" distribution and spreads to involve the fore

head and chin Subvarieties of the rash occur, and of these the 'nevus multiplex of Pringle is perhaps the most frequent Other cutaneous anomalies of less regular occurrence include pendunculated fibromas the so called 'peau de chagrin and *cafe au lait* spots similar to those of von Recklinghausen s disease Retinal lesions though uncommon are of importance in diagnosis when they occur These take the form of grayish-white plaques usually rounded or ovoid, and may be single or multiple Many congenital malformations occur in conjunction with tuberous sclerosis the chief of which include spina bifida cryptorchidism syndactylism and supernumerary digits to mention but a few

Tuberous sclerosis was first described as a clinical syndrome by Bourneville in 1880 The condition derives its name from the potato like nodules which occur in the brain and comprise the basic pathologic lesions of the disease On gross inspection the masses appear smooth and pearly white



FIG 52 Tuberous Sclerosis The tables in the poster or parietal region are increased in width and density and show obliteration of the diploe There are calcific plaques in the superior parietal region the plaques are located intracerebrally

The nodules are hard sclerotic patches which vary widely in size number and distribution When the lesions project into the lateral ventricles they resemble the gutterings of a wax candle Calcification of the nodules is of common occurrence particularly in the region of the lateral ventricles Tuberous sclerosis is a rare disease of protean manifestations which involves primarily the skin and nervous system It is one of the group of congenital ectodermatoses or so-called neurocutaneous syndromes among which are included von Recklinghausen s neurofibromatosis von Hippel-Lindau s disease and angiomatosis cerebri or encephalotrigeminal angiomatosis the so called Sturge-Weber syndrome As the various members of the group of diseases have a common origin in the ectoblast it is not unusual to find varying degrees of overlapping with many common features in these entities

Tuberous sclerosis is rare in North America being more prevalent among the poorer classes of Europe and Australia While generally considered a dysplasia of the ectodermal elements tumors may also occur in regions

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derived from the mesoderm Because of this some observers have considered the disease neoplastic in origin While the anatomical changes appear to date back to the early stages of intrauterine life, they do not terminate at birth Although preformed lesions are capable of continued growth there is probably a long latent period in some instances as some foci may not manifest themselves until puberty or adult life Cases have been reported during the sixth and seventh decades While the disease has definite hereditary tendencies it may also occur sporadically

The principal manifestations comprise epilepsy mental retardation and adenoma sebaceum Other frequent concommitants are congenital tumors of the retina the so called phacomas, subungual fibromas and congenital stigmata such as high arched palate cleft palate simian hands, and multiple skin nodules similar to those in neurofibromata There occasionally occur visceral tumors the most common being rhabdomyoma hypernephroma, renal liposarcoma, and multiple polyposis of the colon In rare instances there is cystic disease of the lung with an irregular diffuse, honeycomb pattern Bone lesions are common in the skull but are unusual elsewhere In mild cases only one or few of the principal manifestations are present Mental deficiency is often recognizable at birth In some instances the infant appears normal only to show deterioration subsequently There is in practically every case delay in walking and talking Retardation of a slight or moderate degree is frequently unrecognized however

Pathology On pathologic examination the disease is characterized by the presence of irregular neoplasms measuring 0 5 cm to 3 0 cm in diameter in the cerebral hemispheres and the walls of the third and lateral ventricles There is involvement of the pyramidal cells and glial elements the pyramidal cells being disoriented atrophied and reduced in number There is marked overgrowth of the neuroglia Giant cells of unknown origin are present in and about the tumors The nodules may undergo cystic degeneration or calcify The nodular lesions in the hypothalamic region are probably the reason for the bouts of fever which may occur in the disease The adenoma sebaceum or nevus multiplex of Pringle comprises a symmetrical butterfly like distribution of small nodular tumors involving the nose cheeks and chin This manifestation usually develops at about the age of four to five years The color is dependent on the degree of vascularity of the tumors and varies from that of the normal skin to a deep red The nodules are composed of hypertrophic sebaceous glands and newly formed connective tissue which may compress and partially or almost entirely replace the glandular elements Skin lesions identical with those of neurofibromatosis such as nodules *café au lait* spots vitiligo and a rough irregular zone of thickened skin on the back known as the peau de chagrin may also occur Studies of the bones at postmortem show various stages of fibrous displacement in the osseous pseudocystic lesions

Roentgen Manifestations The skeletal changes are variable There may be periosteal thickening involving the metacarpals the metatarsal bones and the associated phalanges with generalized fragmentation of the cortical layers In the cranial bones there are diffuse irregular mottling and hazy islands of increased density alternating with areas of rarefaction The inner and outer tables are increased in width and exostoses of the inner table may be present Dickerson describes patchy zones of increased density with areas of osteosclerosis in the skull The marrow spaces of the

cancellous bone in the diploic area are practically completely obliterated in the areas of sclerosis Ackerman has reported a case with cystic changes in the lungs, extensive erosion of a lumbar vertebra, marked rarefaction of the iliac bones with both large areas of increased radiance and small cyst-like formations, and multiple cysts in the necks and juxtatrochanteric regions of the femur A narrow zone of sclerosis surrounded the cyst areas There was fibrillation of the cortex and thickening of the bone trabeculæ in the diaphyses of the femurs In a case recorded by Budenz, there was thickening and notching of several of the ribs similar to that in coarctation of the aorta The calvaria was increased in thickness with widening of the diploe Both the inner and outer tables were irregular and there were multiple exostoses of the inner table in the frontal region both to the right and left of the midline There were irregularly scattered areas of increased density in the occipital region No intracranial calcific depositions were demonstrable The diaphyses of the humeruses and femurs presented increased width and density, irregularity of outline, and coarseness of the trabeculæ A rounded tumor 3 5 cm in diameter was present in the mediastinum adjacent to the antero lateral surface of the fifth dorsal vertebra without erosion of the vertebral border Among the commonest manifestations of tuberous sclerosis is intracranial calcification The basal ganglia are frequently affected In some instances, the calcification may involve the dentate nuclei of the cerebellum and rarely there may be wavy, angiomatous calcific depositions in the occipital pole as in the Sturge-Weber syndrome Some of the areas of apparent intracranial calcification in tuberous sclerosis actually represent sclerotic plaques in the cranial vault In certain instances the skull lesions comprise localized hyperostoses of the inner table of the vault in the region directly adjacent to the nodules in the brain

Lesions in the bones of the hands and feet have been reported Holt and Dickerson describe the osseous changes as cyst-like foci in the phalanges and a distinctive type of periosteal new bone formation along the shafts of the metacarpals and metatarsals The cystic areas are more frequent in the hands while the periosteal changes show a predilection for the metatarsals The cyst like lesions are apparently the result of a slow, progressive rearrangement of the trabeculæ with gradual coalescence and are not sharply defined in character These defects develop during early childhood or in adult life and show a gradual increase in number and size with the passage of a period of years It is believed that the rarefied areas are the result of nonspecific fibrous replacement rather than true cyst formations and are similar in character to the transitory cortical lesions which occur in growing tubular bones and the cystic varieties of neurofibromatosis The periosteal changes comprise a wavy irregular thickening or a localized area of increased density limited to a small segment of the shaft In isolated instances skeletal anomalies similar to those described in neurofibromatosis have also been described in tuberous sclerosis

Differential Diagnosis The osseous lesions may be confused with those of sarcoidosis A fine reticular pattern of bone destruction is characteristically found in sarcoidosis there is less distortion of trabecular pattern and periosteal thickening is very rare entire phalanges being destroyed in many instances without subperiosteal new bone formation In tuberous sclerosis the phalangeal lesions in practically all instances are associated with cranial plaques or intracranial calcifications Chronic arthritis with associated degenerative cystic lesions is differentiated by the character-

istic alterations in the joints The callus formation in healing fractures may cause confusion in diagnosis

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PROGRESSIVE DIAPHYSEAL DYSPLASIA—ENGELMANN'S DISEASE

Progressive diaphyseal dysplasia is also known as Engelmann s disease, periostitis hyperplastica symmetric sclerotic hyperostosis hereditary symmetric osteitis or osteopathia hyperostotica scleroticans multiplex *infantilis* The first case was described in 1929 by Engelmann and only 9 cases had been recorded until 1951 The condition is characterized by symmetrical thickening involving the shafts of the bones resulting in a fusiform enlargement of the diaphyses The cortex becomes thickened by endosteal and periosteal accretion of mottled new bone in which there is absence of recognizable trabecular pattern The affected bone becomes fusiform in shape The lesions are bilaterally symmetrical and are abruptly demarcated The process is slowly progressive extending gradually both proximally and distally along the shaft The epiphyses and the metaphyses are not affected The affected extremities show a relative increase in length with respect to the size of the patient The skull has been affected in two of the recorded cases with amorphous increase in density of the base The soft tissues of the involved extremities show changes similar to those in underdevelopment of the muscles

Clinical Manifestations The symptoms associated with progressive diaphyseal dysplasia comprise vague pains in the limbs a peculiar waddling gait poor appetite and failure to gain weight Various minor abnormalities of the nervous and endocrine system may occur There are no laboratory data which are of significance The patients are of normal intelligence The cause of the anomaly is unknown The condition is self limited There may be a moderate anemia Some of the patients show retardation of growth There is marked muscle atrophy Because of severe muscular underdevelopment and weakness the process appears similar in many respects to muscular dystrophy The ages of the patients have ranged from about two years to twenty-four years It is a lesion which involves the growing bone hence is characteristically seen in younger individuals The symptoms usually develop in the first to the sixth year of life

Roentgen Manifestations There is symmetrical increase in density and thickening of the diaphyses The process affects the compact bones of the shafts the bones being widened the surfaces slightly irregular and the medullary cavities narrow A number of small cystic areas are present in the compact bones The ends of the bones may be normal the epiphyses and metaphyses remaining free of disease, and in older patients the epiphyses unite The changes involve particularly the humeruses and the femurs The radius ulna tibia and fibula on both sides may show similar changes but in some instances are normal in their distal third In the skull there

are sclerotic patches distributed irregularly particularly in the frontal and occipital regions The ribs are large The facial bones clavicles, and other bones of the skeleton may or may not be involved In Engelmann s patient, a boy of eight, there was thickening and sclerosis of the diaphyses of all the long bones and thickened patches in the skull especially in the frontal bones and the bones of the anterior and middle cranial fossas There was involvement of the long tubular bones bilaterally The changes consisted of fusiform enlargement with cortical sclerosis and hyperostosis interspersed with areas of narrowing The petrous portions of the temporal bones were sclerotic In the affected extremities the normal layer of subcutaneous fat was practically absent and the musculature was not decreased in size The changes did not tend to regress rather they became more pronounced with the progressive bone growth of the patient In a case reported by Gillespie and Mussey, the patient was thirty three months of age one of the youngest reported with this disease The syndrome was characterized by progressive symmetrical enlargement and cortical thickening involving the diaphyses of the long bones The changes were present in all the long bones the skull the clavicles and possibly also the ribs In observations made at successive intervals during a period of one year no significant changes appeared in the femurs although there was slight progression of the process in the tibias and fibulas

Pathology Biopsy of the affected bone reveals thickening of the periosteum and sclerosis of the cortex The bone trabeculae are coarse dense and hypertrophied There is increase in the fibrous component of the periosteum The changes are not specific in character The cortex is altered from a compact to a cancellous type the result of combined resorption and deposition of bone associated with remodeling The marrow is altered from a loose fatty type to a fibrous tissue with collections of mononuclear cells The blood vessel walls show thickening

Differential Diagnosis In Engelmann s disease the changes differ from the fibrous dysplasia of bone of the polyostotic type described by Allbright and Lichtenstein and Jaffee in several important respects The osseous changes are not predominantly unilateral in distribution They are characterized by subperiosteal new bone while in fibrous dysplasia this is never seen without associated fractures Pathologically there is no replacement of bone by dense fibrous tissue except that in cases with involvement of the calvaria the changes in the skull are similar to those in polyostotic fibrous dysplasia The lesions are not analogous to the conversion abnormalities of enchondromatosis and Ollier s disease in that previously normal bone precedes the lesion the condition is progressive and there is no cartilage present on pathologic study The changes are similar to the underlying disease process of Paget s disease in that the cortical architecture becomes progressively altered However the process differs from Paget s disease in that radiologically there is no coarse accentuation of the trabecular pattern along the weight bearing lines The new bone is characterized by absence of radiologic pattern and an irregularly amorphous increase in density The serum alkaline phosphatase is normal Histopathologic study does not show the pathognomic mosaic pattern of Paget s disease Differentiation from infantile cortical hyperostosis is more difficult as the microscopic appearance of the two conditions is somewhat similar In progressive diaphyseal dysplasia there is a difference in distribution as infantile cortical hyperostosis shows preference for the mandible, ribs and clavicle and asymmetry

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normalities of the skull and facial bones similar to those in polyostotic fibrous dysplasia may occur and there may also be failure of pneumatization of the temporal bones and nasal accessory sinuses as described by Hermel, Gershon Cohen and Jones The bones heal normally after biopsy The periosteum appears normal but the cortex offers lessened resistance at surgical exploration

Roentgen Manifestations There is *symmetrical and often marked expansion of the metaphyses of the long bones* The changes occur in the femurs tibias fibulas humeruses radiuses, ulnas the bones of the hand the clavicles and the distal aspects of the ribs There is failure of tubulation

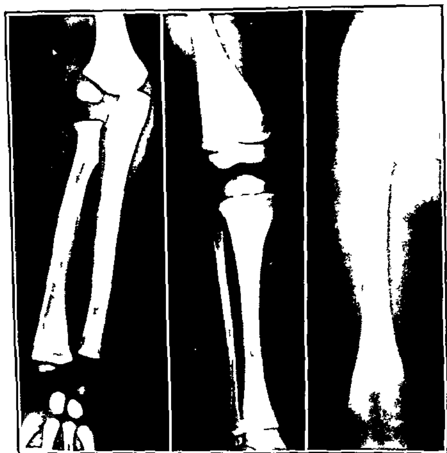


Fig 53 Metaphyseal Dysplasia Pyle s Disease *A* Right forearm *B* Right leg *C* Left femur The metaphyseal portions of the bones are flared and show marked cortical thinning There are multiple transverse striations in the lower femur and upper tibia

of the long bones actual increase in the length of the long bones and widening of the metaphyseal ends of these bones producing a wine-bottle or Erlenmeyer flask appearance In the diseased areas there is cortical thinning osteoporosis and an interlacing trabecular formation The bony outlines and articular surfaces are sharply defined and smooth Osteomalacia is present and the bones are curved with in some instances pathological fractures There may be transverse lines in the bones of the lower extremities In the mid portions of the shafts of the long bones there may be cortical thickening increased density and narrowing of the medullary cavity Ellis in 1934 reported two cases in children presenting an almost identical appearance both clinically and radiologically and emphasized the uniformity within the family group His patients presented clubbing of

of bone involvement Engelmann's disease is progressive in nature, whereas infantile cortical hyperostosis appears to be self limited The disease should be distinguished from the diaphyseal dysplasia (of Caffey) in that no hereditary influences are evident The changes are similar to those which occur in the hereditary multiple diaphyseal sclerosis described by Ribbing However, histologic study reveals no increased bone absorption such as occurs in progressive diaphyseal dysplasia

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FAMILIAL METAPHYSEAL DYSPLASIA PYLES DISEASE SYMMETRICAL SPLAYING OF THE LONG BONES

Familial metaphyseal dysplasia is among the rarest of the generalized disease of bone only four examples of this condition having been recorded in the literature The first case was presented by Pyle in 1931 In 1937 Bakwin and Krida observed the patient reported by Pyle and found the same anomaly of the bones in the patient s sister an albino three years younger Clinically the condition is manifested by knock knees weakness of the feet palpable expansion of the humeruses and the femurs and increased length of the thighs and legs The stature is increased in consequence of elongation of the tibias and femurs Because of the generalized nature of the changes and the occurrence of similar manifestations in siblings the disease is considered familial In the previously recorded cases the parents were normal and other relatives gave no history of bone disease The condition manifests itself early in life knock knee having been noted at the age of eighteen months in one of the patients and fully developed bone changes having been found by Pyle when the patient was three years of age In a case seen at the Boston City Hospital characteristic splaying and rarefaction of the long bones were present in a child eighteen months old The condition is not progressive and does not appear to interfere with the affected individual s general health There are no characteristic alterations in the structure or the chemistry of the blood

The morbid process remains obscure Examination by Ingalls of the femur of a cadaver with a condition apparently identical with the disease under consideration showed a short medullary cavity surrounded by unusually dense bone In the affected portions of the bone the cortex was thin and the cancellous bone was greatly increased in amount and distributed irregularly There appears to be an error or abnormality of the process of tubulation of the long bones which results in marked flaring of the metaphyses thinning of the cortex and elongation of the tubular bones Tubulation is considered a function of the metaphyses Hence the disease is a primary dysplasia of the metaphyseal ends of the long bones Large numbers of transverse lines are present in the bones of the lower extremities indicating that these transverse linear densities represent a manifestation of disordered growth and are not due merely to acceleration or retardation of growth as was formerly believed Osteomalacia is an important concomitant and pathological fractures are common Ab

ing with a new syndrome Soon thereafter several other observers reported similar cases There is no clue as to the etiology of the lesion The entity is frequently confused with other diseases The initial symptom is usually irritability This is often but not in all instances associated with fever There soon develop firm, tender, non pitting deep seated swellings which appear to be situated in the soft tissues The swellings are diffuse and brawny but are not hot or red There is no enlargement of the glands and the lesions do not fluctuate Depositions of subperiosteal new bone develop both in the areas of swelling and also in other regions which are demonstrable only by roentgen examination The deposits may be too small to be palpable or may be very extensive and result in marked enlargement of the affected parts They have been described throughout the skeleton except the phalanges, the small bones of the carpus and tarsus, the vertebrae and the pelvis There may be anemia elevated white count, increased sedimentation rate fever and pleurisy, indicating the



Fig 54 Infantile Cortical Hyperostosis A Roentgenogram of the chest Both clavicles *show increase in width and density with extensive new bone formation B Mandible* There is extensive new bone formation along the inferior aspect of the mandible with irregularity of outline and cortical thickening

possibility that a virus is the causative factor When pleurisy is present it frequently is associated with rib involvement The disease pursues a benign course It is not affected by therapy and in most instances resolves spontaneously being characterized by remissions and exacerbations When multiple lesions are present they vary in appearance and in the stage of development The lesions may regress in one part of the body while advancing in another In no reported case has there been any manifestation of the disease in the parents The tuberculin and serologic tests are negative There is no evidence of rickets or scurvy and therapy for these diseases does not alter its course Irradiation, antibiotics transfusions and antihistamine drugs are ineffective also

While the condition is a definite entity it is not certain whether it represents a new disease It is frequently confused with syphilis osteochondritis or periostitis There is no sex preference Neither race nor geographical location are important the disease having been reported in Holland Germany Canada and throughout the United States The disease occurs in the white the Negro and the Chinese Cases have been present in more than one member of the family but usually not simultaneously It is not

the long bones, neurological symptoms with optic atrophy and bilateral sixth nerve palsies and a peculiar facies and cranial conformation Roentgen study revealed thickening and sclerosis of the middle thirds of the long bones flaring of the metaphyses stippling of the epiphyses and increased density at the base of the skull

Differential Diagnosis Many other aberrations of growth produce similar abnormalities and it appears that analagous patterns may be produced by a variety of causes Gaucher s disease presents a failure of tubulation with an Erlenmeyer flask appearance as in *Pyle's disease* Underconstriction of the long bones may occur in diseases which infiltrate the bone marrow and has been described in leukemia and erythroblastic anemia in deficiency states such as healing rickets and scurvy and cystic fibrosis of the pancreas and in developmental diseases including chondrodystrophy and osteopetrosis Engelmann s disease progressive diaphyseal dysplasia is characterized by widening and hyperostosis of the diaphysis with fusiform widening and distortion of the mid shafts of the long bones the metaphyses not presenting the changes found in familial metaphyseal dysplasia

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HEREDITARY MULTIPLE DIAPHYSEAL SCLEROSIS (RIBBING)

Hereditary multiple diaphyseal sclerosis was described by Ribbing in 1949 The disease is characterized by fusiform thickening of a portion of the diaphysis of a long bone There is marked density of the affected segment of the bone with practically complete obliteration of the cancellous portion The changes tend to be multiple and symmetrical The tibia and femur are the most common sites The ends of the shafts of the bones appear essentially normal There may be local pain in some instances Frequently there are no clinical manifestations and the condition is discovered only on roentgen study The cause of the condition is unknown The changes progress for a few years then become stationary The disease has been reported only after puberty The time of onset of the condition has not been established Engelmann s disease occurs at an earlier age and is part of a more widespread disorder with disturbances of gait and posture and retardation of growth in height and weight In hereditary multiple diaphyseal sclerosis there is only new bone formation while in Engelmann s disease there is new bone deposition associated with progressive absorption

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RIBBING S Hereditary Multiple Diaphyseal Sclerosis Acta Radiol 31 522 1949

INFANTILE CORTICAL HYPEROSTOSIS

This condition was first recognized in 1930 by Roske who described a male infant with painful deposits of subperiosteal new bone in the left mandible and the humerus tibia and radius bilaterally He concluded that he was deal

after the fourth month of life Despite the fact that the disease is relatively benign in character, it is imperative that the syndrome be recognized because of the severity of other diseases which must be included in differential diagnosis The painful extremities in conjunction with the other symptoms suggest scurvy, chronic vitamin A poisoning, poliomyelitis, leukemia, rheumatic fever, rheumatoid arthritis and osteomyelitis The facial swelling has caused confusion with infectious parotitis, osteomyelitis, neoplasm of the mandible, and traumatic injury In addition, similar cortical thickenings have been noted in syphilis and rickets In the establishment of the diagnosis of infantile cortical hyperostosis, age is a decisive factor The average age of onset of the disease is nine weeks, and onset after the age of five months constitutes a valid cause for questioning the diagnosis

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SHUMAN H H Infantile Cortical Hyperostoses J Ped *32* 195 1948

SIDBURY J B JR and SIDBURY J B Infantile Cortical Hyperostoses N E J Med *250* 309 1954

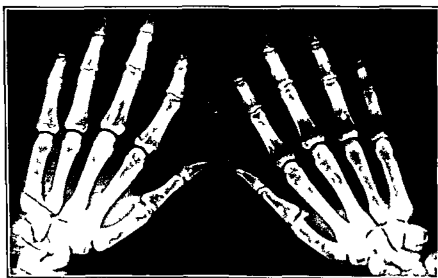


FIG 55 Osteopoikilosis There are multiple small rounded areas of increased density within the bones The involvement is very extensive similar densities being present in the upper femurs and the bones of both feet

OSTEOPOIKOLOSIS OSTEOPATHIA CONDENSANS DISSEMINATA

Osteopoikilosis is an unusual lesion of the skeleton It is characterized by the presence of multiple small areas of increased density irregularly distributed in many bones usually in the cancellous portions The dense spots are circular ovoid or oblong and usually lie with the long axis parallel to the long axis of the affected bone They vary in size from 2 0 mm to 3 0 cm or more frequently occur in the epiphysis and adjacent parts of the metaphysis usually being more numerous in the latter and have been described in the bones of the knee including the patella and the bones of the carpus and the tarsus Involvement has been noted in every bone of the body with the exception of the skull, the ribs, and the vertebræ

related to rickets, scurvy, syphilis, tuberculosis pyogenic infection, allergy or trauma It does not appear to be a disorder of mineral metabolism or a disturbance of bodily metabolism The treatment is entirely symptomatic and supportive Penicillin and sulfonamide preparations have not proven of value Sidbury and Sidbury consider it the clinical manifestation of an inherited defect of the arterioles supplying the affected area, the resulting hypoxia causing focal necrosis of the overlying soft tissues and proliferative reaction of the periosteum The evidence that infection, trauma, allergy, endocrinopathy, maternal causes birth factors and dietary aberrations are etiologic agents is slight and they emphasize the fact that the underlying fault may be an intrauterine or genetic factor

Caffey and Silverman in 1944 published a description of the syndrome and entitled it "Infantile Cortical Hyperostoses" The principal features of the disorder as described by them include onset during the early part of the first year of life, tender swelling in the face the jaw, the scapular regions and the extremities and multiple scattered hyperostoses demonstrated by roentgen study in bones adjacent to the tender swellings and also in other bones whose overlying soft tissues appear normal on clinical and roentgen examination Biopsies of the affected bone show hyperplasia of the lamellar cortical bone The swellings over the sites of the bone involvement tend to progress rapidly and regress slowly Smyth *et al* describe a series of seven cases appearing in infants and sometimes in older children Their cases were characterized by irritability fever, anemia leukocytosis and a periosteal reaction in numerous bones In their patients the constant finding was a periosteal reaction characterized by new bone formation with a laminated onion peel appearance Biopsies of the affected bones and other clinical studies were of no aid in determining the etiology Biopsies of the affected muscles in two cases showed evidence of dystrophy suggesting a metabolic factor All the patients survived and in most instances the condition resolved spontaneously The lesion is not as rare as was formerly supposed

Roentgen Manifestations The changes are most common in the mandible clavicle scapula and tubular bones including the metatarsals and comprise a periosteal reaction or hyperostosis of the cortices In the case reported by Whipple there was involvement of the skull and the bones of the calvaria Pleurisy may occur in association with involvement of the ribs Characteristically there is thickening of the external cortical portion of the bone with subperiosteal new bone formation along the diaphysis This may be lamellated and involves the entire length of the corticalis except for the terminal segments of the shafts There is extensive subperiosteal new bone formation The changes develop rapidly and undergo slow regression The course is usually prolonged the disease as a rule persisting for many months The process eventually resolve completely and leaves no demonstrable residuals in most instances

The diagnosis is dependent on the clinical manifestations of fever irritability anemia and brawny facial edema and the roentgenographic evidence of hyperostosis of the bones The administration of antihistamine preparations is ineffective There is no alteration in the growth or development of the patient throughout the course of the disease A characteristic of the syndrome is the onset in the first few weeks or months of life Caffey states that two older patients described by him in 1946 actually suffered from excessive vitamin A intake rather than infantile cortical hyperostosis and expressed the opinion that no true case begins

intermittent defect of this type gives rise to a persistent structureless area of density in the bone The area is rounded and is characterized by an abrupt transition from normal, apparently healthy bone to complete absence of reticulation in the affected zone When diffuse, widespread and abundant, the process is called osteopoikilosis The lesions appear innocuous Cases have been reported in which hypopituitarism was present as a complicating factor The disease seems to have hereditary tendencies The blood calcium and phosphorus levels are normal There are no symptoms, the condition being discovered only by roentgen examination In the differential diagnosis, it is necessary to consider osteoblastic metastatic carcinoma, bone infarct and medullary osteoma There is no therapy The prognosis is good

ADDITIONAL READING

HIRSCH I S Osteopoikilosis Radiol 25 349 1935
THOMPSON R H HOOVER R and FULTON H F Osteopoikilosis Am J Roent 49 603 1943

MULTIPLE EPIPHYSEAL DYSPLASIA DYSPLASIA EPIPHYSIALIS MULTIPLEX

Dysplasia epiphysialis multiplex is a rare congenital developmental anomaly characterized by mottling and irregularity of outline of the epiphyses, dwarfism and shortening of the digits The first instance of this syndrome was reported in 1934 by Jansen and he termed it epiphyseal dysostosis Fairbank in 1935 described the same condition and gave it the name dysplasia epiphysialis multiplex It occurs in both sexes, more frequently in males and does not appear to be hereditary or familial The cause is not known The changes may be limited to a few or involve many of the epiphyses It is most common in the hips, shoulders ankles, knees, wrists and elbows In addition to the changes in the epiphyses there may be irregularity of the outline of the heads of the metacarpals, the metatarsals and the tarsal and carpal bones Clinically the condition is manifested by difficulty in locomotion due to pain and stiffness of the joints There are varying degrees of dwarfism The epiphyses may appear enlarged The digits are short Flexion deformities of the knees and elbows and laxity of the wrists and knees may be present Shortening and widening of the vertebral bodies has been described

Roentgen Manifestations The principal abnormalities are in the epiphyses The centers of ossification of the epiphyses are late in appearance slow in development and delayed in fusing with the shaft There is marked irregularity of ossification The epiphyseal centers are irregular mottled and discrete The stippling of the epiphyses is less marked than in punctate epiphyseal dysplasia The affected epiphyses tend to become normal in density with the passage of time although the outlines remain permanently irregular The most characteristic changes occur in the ankles The lower epiphysis of the tibia is diminished in size and the articular surface of the inferior aspect of the tibia is tilted The astragalus also undergoes tilting and is altered in shape to conform with the deformity of the tibia The fibula may be elongated and the external malleolus lies

However even in these bones one or two spots may occur the skull being the least common site The areas of density lie in the cancellous bone and as a rule are entirely free from the cortex The cortex is not distorted or otherwise altered in any way In some instances the area of increased density assumes the form of a short streak parallel with the long axis of a



Fig 56 Osteopoikilosis *A* Pelvis and hips *B* Left knee anteroposterior view *C* Left knee lateral view There are numerous small rounded areas of increased density within the bones about the hips and knees The areas are rounded or oval in shape vary in size and are sharply defined

tubular bone particularly when the lesion is situated some distance from an epiphyseal line of a major long bone The areas of density are seldom found in the shaft Patients have been observed over a number of years with no apparent change in the roentgen manifestation

It is believed that defective bone formation in the metaphysis results in the formation of dense non reticulated calcareous depositions A central

intermittent defect of this type gives rise to a persistent structureless area of density in the bone The area is rounded and is characterized by an abrupt transition from normal, apparently healthy bone to complete absence of reticulation in the affected zone When diffuse, widespread and abundant, the process is called osteopoikilosis The lesions appear innocuous Cases have been reported in which hypopituitarism was present as a complicating factor The disease seems to have hereditary tendencies The blood calcium and phosphorus levels are normal There are no symptoms, the condition being discovered only by roentgen examination In the differential diagnosis, it is necessary to consider osteoblastic metastatic carcinoma, bone infarct and medullary osteoma There is no therapy The prognosis is good

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in an abnormally low position Similar changes have not been described in the radius and ulna although in one case reported by Wiles, the radius appeared relatively long The metaphyses may show flaring The shafts of the long bones may be short and widened The carpal and tarsal bones ossify late and are markedly irregular in outline The metacarpals metatarsals and phalanges are short and there is irregular ossification of the epiphyses No changes have been described in the skull and teeth

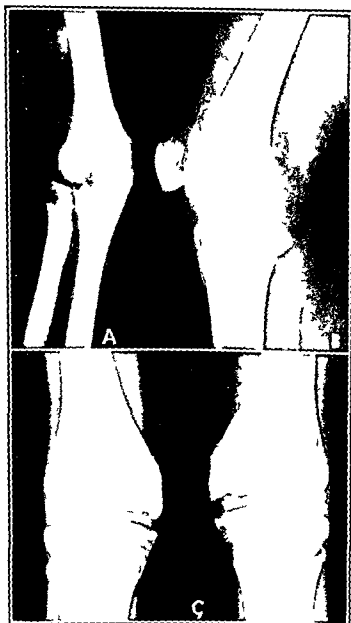


FIG 57 Dysplasia Epiphysialis Multiplex *A* Left elbow The elbow joint space is irregular in outline There is marked irregularity of outline of the articular surfaces of the bones of the elbow The soft tissues about the elbow are thickened *B* Right knee lateral projection There is irregularity of outline involving the articular surfaces of the lower end of the femur the upper end of the tibia and the posterior aspect of the patella The space between the patella and the femur is narrowed *C* Knees anteroposterior projection The articular surfaces of the bones of the knee are narrowed and irregular in outline and show multiple shallow areas of increased radiance The knee joint spaces are decreased in width The soft tissues about the knees are thickened

TARSO EPIPHYSEAL ACLASIS

Tarso epiphyseal aclasis is an unusual developmental disorder of ossification centers which is manifested by eccentric cartilaginous overgrowth with consequent asymmetrical enlargement of the involved ossification centers It may affect any of the osseous structures of the lower limb including the tarsals and the bones of the foot The first report of this condition was made in 1926 by Mouchet and Belot who published the case of an 8 month old boy with involvement of the tibia and astragalus They termed the condition "La Tarsomegalie" Their patient was observed until he reached the age of 30 months Trevor in 1950 reported a series of ten patients with this abnormality He demonstrated the characteristic distribution of the lesion and suggested the name "tarso epiphyseal aclasis' Geyman in 1931 reported an unusual case and the reproductions of his radiographs show changes in the knee and ankle which are highly suggestive of this anomaly However, the clinical findings and description of skeletal involvement elsewhere in the body do not coincide with the entity as described by Trevor

Nature and Distribution of the Lesion The abnormal ossification centers show eccentric overgrowth as the chondromatous portion enlarges asymmetrically Diffuse amorphous calcific stippling and/or ossification is

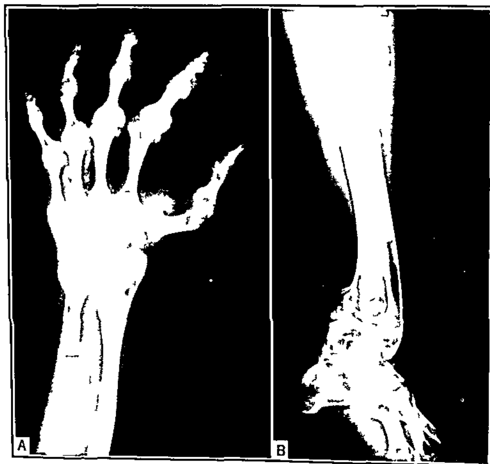


Fig 58 Multiple Epiphyseal Dysplasia *A* Left wrist and hand The articular surface of the lower end of the radius is irregular in outline and concave The fingers are short *B* Lower leg and ankle The tibia is short and its inferior articular surface is tilted and irregular in outline The talus is irregular and tilted and its shape is altered to conform to the tibial deformity The fibula is elongated and bowed The external malleolus lies in an unusually low position

scattered throughout the anomalous cartilaginous mass. An apparent characteristic feature is noted when more than one epiphysis is involved in which instance all of the epiphyseal centers affected show overgrowth of either the medial or lateral portions. Thus, one limb may show involvement of the medial femoral and tibial epiphyses at the knee and the medial malleolus at the ankle. None of the reported cases have varied from this. In a case in which both lower limbs were involved, the medial condyle of the femur on the left and the medial aspects of the "ankle and foot" on the right were affected. However, in case of involvement of the bones of the foot this pattern is not invariable.

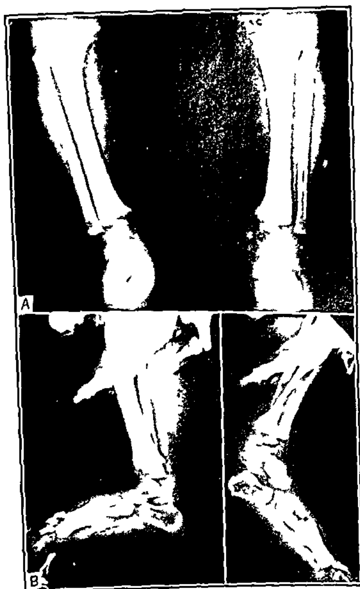


Fig. 59. Tarso Epiphyseal Aclasis. A. Lower legs antero posterior projection. Age eight months. The ossification centers of the lower end of the left tibia and the astragalus are enlarged asymmetrically. There is irregular calcification within the overgrown cartilaginous portions. The nucleus of the distal tibial epiphysis is larger than that on the right. The upper tibial epiphyses are symmetrical and appear normal in size and contour. *B*. Lower legs lateral projection. Age eight months. The enlarged cartilaginous portions of the left astragalus and tibia are irregularly calcified. There is anterior displacement of the left astragalus with relation to the tibia. The left tibia shows metaphyseal widening concomitant with the epiphyseal overgrowth, however the distal portion of the diaphysis is well modelled and of normal size. Several small irregular ossific nuclei indicate the precocious appearance of the internal cuneiform on the left.

The bones most frequently affected were the ossification centers of the distal femur the astragalus the distal tibia and the proximal tibia The patients exhibited no evidence of disease or associated anomaly other than the osseous abnormality In all the patients the deformities were muscular and/or osseous and were apparently the result of the underlying bone and cartilaginous overgrowth and consequent faulty joint mechanics Growth and development, both physical and mental, were unimpaired It is a noteworthy fact that of eleven recorded cases of this disease, ten were in males

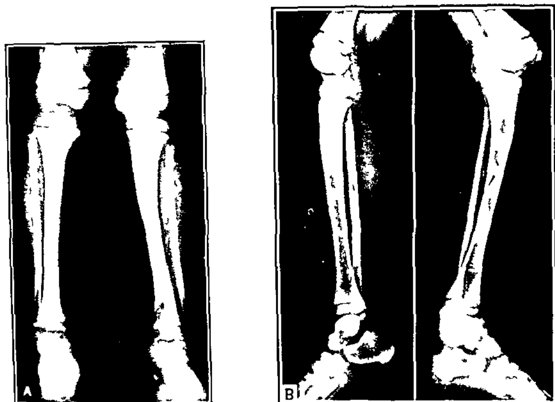


FIG 60 Tarso Epiphyseal Aclasis Same patient as in Figure 59 A Lower legs antero posterior views Age four years ten months The bony and cartilaginous overgrowths have progressed particularly in the region of the knee where there are well defined bony masses with peripheral corticalis and central trabecular pattern The overgrowths are in relation to the medial aspects of the left lower femoral and upper tibial epiphyses The extensive involvement of the lower tibial epiphysis medially has resulted in definite enlargement of the internal malleolus The deformed astragalus is approximately twice normal size The left genu valgum fibular and muscular under development and osteoporosis are present as previously *B* Lower legs lateral views Age four years ten months The ossific nucleus of the left patella is larger than on the right The cartilaginous ossification centers show no discrepancy in size The bony masses at the knee extend posteriorly into the popliteal space The left astragalus shows massive overgrowth and gross distortion of outline The adjacent articular surfaces of the os calcis are unaffected

Clinical Manifestations The parents are first made aware of the abnormality by mechanical interference with joint function and/or the presence of a mass There is no pain the patients appearing healthy, active and normal both physically and mentally

Roentgenographic Findings Initially there is amorphous calcific stippling in the asymmetrically enlarged portions of the affected ossification centers As the child grows older the calcification becomes more extensive and irregular ossification takes place When more than one epiphysis is involved the changes are limited to either the medial or lateral

scattered throughout the anomalous cartilaginous mass. An apparent characteristic feature is noted when more than one epiphysis is involved in which instance all of the epiphyseal centers affected show overgrowth of either the medial or lateral portions. Thus, one limb may show involvement of the medial femoral and tibial epiphyses at the knee and the medial malleolus at the ankle. None of the reported cases have varied from this. In a case in which both lower limbs were involved the medial condyle of the femur on the left and the medial aspects of the "ankle and foot" on the right were affected. However in case of involvement of the bones of the foot, this pattern is not invariable.

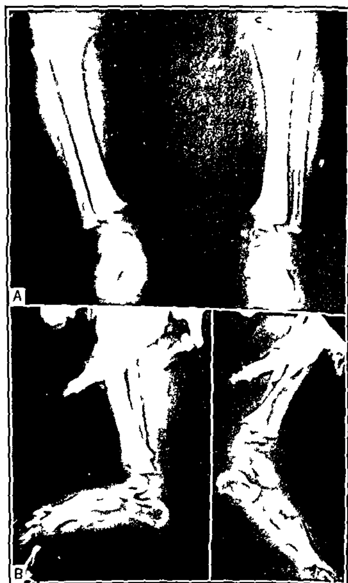


FIG 59. Tarso Epiphyseal Aclasis. *A* Lower legs antero posterior projection. Age eight months. The ossification centers of the lower end of the left tibia and the astragalus are enlarged asymmetrically. There is irregular calcification within the overgrown cartilaginous portions. The nucleus of the distal tibial epiphysis is larger than that on the right. The upper tibial epiphyses are symmetrical and appear normal in size and contour. *B* Lower legs lateral projection. Age *eight months*. The *enlarged* cartilaginous portions of the left astragalus and *tibia* are irregularly calcified. There is anterior displacement of the left astragalus with relation to the tibia. The left tibia shows metaphyseal widening concomitant with the epiphyseal overgrowth, however the distal portion of the diaphysis is well modelled and of normal size. Several small irregular ossific nuclei indicate the precocious appearance of the internal cuneiform on the left.

aspect of the limb, e g, the lateral femoral and tibial condyles and the lateral malleolus at the ankle The bony enlargement may extend anteriorly or posteriorly When the bones of the foot are involved, the asymmetrical pattern of the changes is less pronounced

Pathogenesis The etiology of tarso epiphyseal aclasis is unknown Laboratory investigations including serologic studies reveal normal findings There is no demonstrable relation to trauma or infection, and no evidence that the changes are neoplastic in character It is evidently developmental in origin, as suggested by Trevor, who postulates a disruption of the normal cellular activity of the cartilaginous ossification center This normally follows an orderly sequence of cell proliferation, maturation, senescence, and disintegration According to Trevor's thesis, cells from one portion of the "mitotic annulus" continue to multiply unduly without following the usual processes of maturation and eventual cell death The term "aclasis' used in the title derives from this failure of cellular breakdown Because of the striking constancy of the pattern of osseous involvement, we have attempted to find an explanation on the basis of the embryology of the lower extremity There is, however, relatively little specific information with regard to the early development of the limb bud The most revealing studies are those of Steiner in the field of comparative embryology His investigations, together with the homologies of the tarsal and carpal bones, help to explain the diffuse involvement of the ossification centers of the bones of the foot No definite correlation is evident between the embryology of the tibia and fibula and the pattern of the lesion in these bones Trevor suggests a possible relationship between the hemi epiphyseal distribution of the dysplasia and the blood supply of the epiphysis According to Harris, nutrient vessels course along the joint capsule enter the nucleus and supply the adjacent portion of the epiphysis, terminating in an anastamotic network In the presence of such a relationship in the local blood supply, it becomes possible to ascribe some of the findings to the effects of local hypervascularity This could account for the premature appearance of ossification centers which often occurs in this disease, whether involved in the dysplastic process or not It also would aid in explaining the diaphyseal elongation which occurred in one patient

There has been no indication of a neoplastic tendency in the hypertrophied chondromatous tissue The available biopsy reports show normal hyaline cartilage with foci of calcification and ossification with no evidences of rapid growth metaplasia or neoplasm formation Nonetheless, cartilage proliferating abnormally should always be viewed with suspicion and the patients should be observed frequently with this potentiality in mind

Differential Diagnosis *Chondrodystrophia calcificans congenita (stippled epiphyses)* In this condition there are scattered foci of calcification distributed throughout affected ossification centers The epiphyses may be enlarged but are symmetrical and regular in outline In tarso-epiphyseal aclasis there is eccentric chondromatous overgrowth the abnormal calcific foci being limited to the enlarged portions of the bone In many cases of stippled epiphyses the diaphyses are short and thick with consequent dwarfing there are flexion contractures at the affected joints often with para articular fibrosis and calcification there is mental dullness, and bilateral congenital cataracts are common None of these are present in tarso epiphyseal aclasis and in a patient seen at our hospital there was

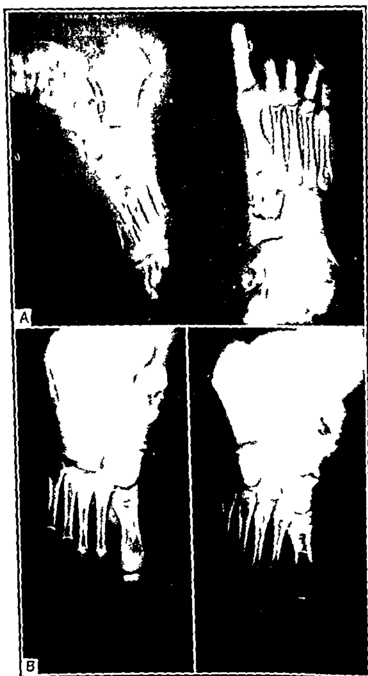


Fig 61 Tarso Epiphyseal Aclasis Same patient as in Figure 60 *A* Left ankle and foot Age five years eleven months All of the ossification centers along the medial apsect of the foot including the terminal phalanx of the great toe are affected They are however at differing stages in the dysplastic process The earliest manifestation is at the epiphysis of the proximal phalanx of the great toe where there is only scattered calcification beyond the normal confines of the ossification center The first metatarsal shows along its dorsomedial aspect the next stage of the disease with a small bony outgrowth which is continuous with the ossific nucleus Late extensive enlargement is present in the internal cuneiform the scaphoid and the astragalus The terminal phalanx of the great toe presents a crescentic bony mass along the lateral margin of the interphalangeal joint *B* Left foot Age seven years one month There is evidence of enlargement of the cartilaginous portion of the epiphysis of the proximal end of the distal phalanx of the great toe and stippling as previously The involvement of the epiphysis of the first metatarsal is more pronounced and the abnormal bony mass along the distal medial margin of the talus is larger than it was fourteen months before

presumably beyond pubescence If the patient can be maintained through the years before puberty with conservative measures it appears that there is favorable prospect for eventual arrest of the developing bony masses Should surgery be necessary it may comprise (1) local excision of the cartilaginous mass (2) excision of the abnormal growths with arthrodesis, and (3) osteotomy and other plastic procedures to correct valgus and varus deformities

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D'ANCIO G J RITVO M and ULIN R Clinical and Roentgen Manifestations of Tarso Epiphyseal Aclasis Am J Roent in press

MOUCHET A and BELOT J La Tarsomegalie J de Rad et D Elect *10* 289 1926

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OSTEOPATHIA STRIATA

Osteopathia striata is a rare condition which is characterized by striation of the skeleton particularly of the metaphyses of the long bones The first report of this condition was published by Voorhoeve Fairbank in 1925 recorded a similar case in a boy of twelve and suggested the title for this condition There are both hereditary and familial influences in osteopathia striata Most of the cases are in males and in young individuals from ten to fifteen years of age The cause of the condition is unknown Fairbank considers that it is the result of a congenital developmental error It has been suggested that the striated bones are related to dyschondroplasia and osteopoikilosis and that the 3 diseases are variants of the same fundamental order with the first forming a link between the other two

Clinical Manifestations There are no symptoms which are directly referable to the condition The patients may complain of vague recurrent joint pains with slight swelling The joints most frequently involved appear to be the hip knee and shoulder In most cases the physical examination is negative and studies of the blood reveal nothing of importance

Roentgen Manifestations The condition is characterized by striations which affect in varying degrees all of the bones except the skull and the clavicles In the long bones the striation is principally in the metaphyses and is characterized by dense lines extending parallel to the long axis of the bones and in some instances for a considerable distance into the shaft The thickness of the striations is widely variable but in most cases they are fine and linear There may be osteoporosis in the portions of the bone between the linear striations In some instances the dense lines appear to extend into the adjacent epiphyses There may be mottling with dense and clear spots in the epiphysis The clavicles do not appear to be affected and the hands are less markedly involved than the remainder of the skeleton The striations are well seen in the tarsus particularly the os calcis The patella may be affected In the ilium the striations form lines which radiate towards the crests similar to the columns of cartilage in dyschondroplasia In a unilateral case described by Fairbank there was a dense patch in the ischium on the side on which the striation on the long bones was most marked with a few dense irregular spots in the opposite ischium Re examination after an interval of a few years showed that the changes had become *more* marked In most instances the cortex of the bones is normal in thickness and density and there is no distortion Small exostoses may be present

diaphyseal lengthening rather than shortening Common to both diseases is the occasional premature appearance of tarsal ossification centers, whether or not involved in the dysplastic process

Chondrodystrophy (*achondroplasia*) In this generalized condition, all the tubular bones are short with broad metaphyses In the hyperplastic form they are often tufted and irregular *Particularly in this group the* ossification centers may show spotty calcification, but there is no overgrowth of the center or other changes resembling the eccentric localized lesion in tarso epiphyseal aclasis

Dyschondroplasia (*Ollier s Disease*) Ollier s disease affects the metaphyses of the long bones and is characterized by columns and islands of proliferating cartilage, some of which undergo extensive calcification In severe cases the distortion is so great that the changes at the epiphyseal plate produce an appearance which might be confused with advanced tarso epiphyseal aclasis However, in dyschondroplasia, there is predominantly metaphyseal involvement with secondary shortening of the diaphysis While the epiphysis may undergo deformation conforming to the metaphyseal contour it does not share in the chondromatous overgrowth and calcification which occur in the adjoining shaft

Dysplasia Epiphysialis Multiplex In dysplasia epiphysialis multiplex the ossification centers appear late and develop slowly They may be enlarged and irregular and present scattered calcific foci throughout their confines The patients usually are short in stature because of the stunting of the diaphyses and may complain of pain and stiffness in the affected joints Differentiation is established by the limited asymmetric changes in tarso epiphyseal aclasis and the absence of pain diaphyseal shortening or delayed skeletal maturation On the contrary tarsal ossification often is premature

Cretinism Many hypothyroid infants show an aberration of skeletal development termed ' cretinoid epiphyseal dysgenesis This is characterized by irregularly ossified epiphyses most commonly those of the proximal femur and humerus While the appearance may be similar to that in the early stages of tarso epiphyseal aclasis the affected epiphyses in the cretin tend to be smaller than normal in size irregular in outline and without eccentric osteochondromatous masses The characteristic facies of the cretin the retarded mental and physical development and dwarfism establish the differentiation between tarso epiphyseal aclasis and cretinism

Aseptic Necrosis Primary aseptic necrosis of the astragalus (Mouchet) tarsal scaphoid (Koehler) medial cuneiform (Buschke) and medial tibial condyle (Blount) affect some of the bones frequently involved in tarso epiphyseal aclasis Points of similarity in the two diseases include irregular ossification and fragmentation of the affected nuclei However painless eccentric epiphyseal overgrowth and calcification the essential features of tarso epiphyseal aclasis are absent in osteochondritis

Calcified Hematoma Amorphous calcification may take place in hematomas When located near a joint it may be confused with a disturbance of epiphysis The calcification is periarticular and not in cartilage continuous with the ossification center as occurs in tarso epiphyseal aclasis

Treatment Conservative management with braces and special shoes suffices The cartilaginous proliferation continues during the early years With the advent of puberty the progression of the disease may be expected to cease This is implied by the eventual arrest of the condition in one of the patients in Trevor s series who was observed for 15 years and therefore

presumably beyond pubescence If the patient can be maintained through the years before puberty with conservative measures, it appears that there is favorable prospect for eventual arrest of the developing bony masses Should surgery be necessary, it may comprise (1) local excision of the cartilaginous mass, (2) excision of the abnormal growths with arthrodesis, and (3) osteotomy and other plastic procedures to correct valgus and varus deformities

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OSTEOPATHIA STRIATA

Osteopathia striata is a rare condition which is characterized by striation of the skeleton particularly of the metaphyses of the long bones The first report of this condition was published by Voorhoeve Fairbank in 1925 recorded a similar case in a boy of twelve and suggested the title for this condition There are both hereditary and familial influences in osteopathia striata Most of the cases are in males and in young individuals from ten to fifteen years of age The cause of the condition is unknown Fairbank considers that it is the result of a congenital developmental error It has been suggested that the striated bones are related to dyschondroplasia and osteopoikilosis and that the 3 diseases are variants of the same fundamental order with the first forming a link between the other two

Clinical Manifestations There are no symptoms which are directly referable to the condition The patients may complain of vague recurrent joint pains with slight swelling The joints most frequently involved appear to be the hip knee and shoulder In most cases the physical examination is negative and studies of the blood reveal nothing of importance

Roentgen Manifestations The condition is characterized by striations which affect in varying degrees all of the bones except the skull and the clavicles In the long bones the striation is principally in the metaphyses and is characterized by dense lines extending parallel to the long axis of the bones and in some instances for a considerable distance into the shaft The thickness of the striations is widely variable but in most cases they are fine and linear There may be osteoporosis in the portions of the bone between the linear striations In some instances the dense lines appear to extend into the adjacent epiphyses There may be mottling with dense and clear spots in the epiphysis The clavicles do not appear to be affected and the hands are less markedly involved than the remainder of the skeleton The striations are well seen in the tarsus, particularly the os calcis The patella may be affected In the ilium the striations form lines which radiate towards the crests similar to the columns of cartilage in dyschondroplasia In a unilateral case described by Fairbank there was a dense patch in the ischium on the side on which the striation on the long bones was most marked with a few dense irregular spots in the opposite ischium Re examination after an interval of a few years showed that the changes had become more marked In most instances the cortex of the bones is normal in thickness and density and there is no distortion Small exostoses may be present

Pathology No microscopic study has been made of any of these cases However, it is believed that the bone is essentially normal The striations of the metaphyses do not appear to be the result of an epiphyseal fault

Differential Diagnosis In dyschondroplasia the bone between the columns of unossified cartilage often gives an appearance which is striated This change is less extensive and occurs with less regularity in the major epiphyses In osteopathia striata the long bones of the hand and foot are less extensively affected than the major long bones, while in dyschondroplasia the hands, and to a lesser extent the feet, are the chief regions which show the striations It must be stressed that there is a fan like arrangement of the striations in the ilium in both disorders In osteopoikilosis the epiphyses particularly and other portions of the skeleton are peppered with dense spots While these spots may be prolonged and in certain areas give an appearance of streaking they are much shorter and less regular than the dense lines which are characteristic of osteopathia striata In melorheostosis, dense streaking may occur but it is limited in extent and usually involves but one or two bones The character and limited distribution of the other changes are atypical In polyostotic fibrous dysplasia patches of increased density ending in 2 or 3 broad streaks may occur These usually involve only one or two bones In neurofibromatosis there may be a suggestion of streaking Generalized hyperostosis with pachydermia affects the long bones and may be associated with coarse striations in portions of the skeleton This condition may occur without alterations in the skin It has been linked with pulmonary hypertrophic osteoarthopathy However the changes are not exactly similar and the diagnosis can usually be established on the basis of the clinical and roentgen manifestations In generalized hyperostosis with pachydermia there are coarse striations of the cancellous bone and thickening of the cortices of the long bones The striation is much coarser than in osteopathia striata and is seen particularly well in the metaphyses and epiphyses of the long bones the vertebræ and the tarsal bones The hyperostosis is usually on the external rather than on the internal surfaces of the cortex and involves the shafts of the long bones The density of the new bone varies considerably In the metacarpals metatarsals and to a lesser degree the phalanges there is no more than increased thickness of the dense cortex In the major long bones the new bone is more variable in density although as a rule it is less dense than the normal cortical bone and has an irregular fluffy surface In some instances the thickened bones present an irregular honeycomb appearance similar to that in Paget s disease The skull ribs and bodies of the vertebræ are usually not affected

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DYSTROPHY OF THE NAILS ANOMALY OF THE PATELLA ARTHRODYSPLASIA CHONDRODYSPLASIA AND ILIAC HORNS TURNER S SYNDROME

There have appeared in the literature isolated reports describing an unusual association of malformation of the nails hypoplasia or total absence of the patellas and deformities with impaired function of the

elbow These anomalies are congenital and are inherited in varying modifications in several generations In certain instances, one or more of the cardinal features of the triad has been absent or modified The pathogenesis apparently lies in a developmental defect in the ectodermal and mesodermal layers of the embryo In 1897 Little reviewed the literature of congenital absence or delayed development of the patellas and added three new cases Mayer also reported a case in the same year In 1900, Wolf recorded an instance in which a mother showed congenital absence of the thumb nails and both patellas Two children, a son and a daughter, inherited the deformities while the grandchildren were normal The Senturias in 1944 described patients with this anomaly and other authors have contributed additional instances

While there is considerable variation in the nature and degree of the congenital defects in the cases recorded in the literature, the localization of the abnormalities in the nails the patellas, and the elbows has been striking In the characteristic case, the disorder of the nails varies from total absence of the nail to one that is only a little thinner than normal It involves the thumb most severely and becomes less marked as one approaches the little finger The affected nail presents a small or normal base, is much thinner than normal, and extends only about half way to the tip of the finger This permits the fleshy end of the finger to turn backward over the nail There is hemiatrophy of the ulnar side of the nail in many instances In other cases there is complete absence of the nail of the thumb with, in some instances, associated involvement of the remaining fingers and occasionally the toes The deformities of the patella are characterized by total absence or severe hypoplasia The knees are flat when extended and square or angular when flexed A hypoplastic patella located on the external aspect of the knee lies at a higher level than usual There is marked prominence of the internal condyle of the femur Genu valgum may be present The involved elbow shows an unusually prominent internal condyle with an increased carrying angle The patient finds it impossible to fully extend the forearm and supination is limited Roentgen study of the elbow reveals an arthrodysplasia which is characterized by elongation deformity and subluxation of the proximal end of the head of the radius The radial head is poorly formed and the superior radioulnar articulation is dysplastic In other instances, there is extensive overgrowth of bone in the region of the elbow with secondary arthritic changes

Other less constant and variable features may occur The ankles may be thick with large malleoli particularly the internal malleolus Prominence of the acromial ends of the clavicles, undersized scapulas, and small coracoid and acromial processes have been observed In the hip and pelvis there may be an increase in the angle between the neck and the shaft of the femur and deepening of concavity of the external surface of the ilium imparting to the crest the appearance of a flare of its posterior half There also have been reported a leaf shaped discoloration of the iris and minor changes in the shoulder girdle There may be a laxity of the metacarpo phalangeal and interphalangeal joints making it impossible for the patient to extend the digits to an angle of 90 degrees without discomfort The so called iliac horns are an unusual anomaly of the pelvis characterized by bony processes which extend bilaterally from the posterior aspects of the iliac bones The iliac horns do not appear to be vestigial structures The condition is asymptomatic and is usually discovered as an incidental finding during roentgen examination The lesion is considered to be on a genetic basis

and constitutes a hereditary arthrodysplasia or familial dyschondroplasia and has been termed Turner s syndrome in the literature The bony protuberances from the iliac bones may occur as the only manifestation or in association with other changes The iliac horns are not a constant feature of the hereditary syndrome in many instances the pelvis being normal while other manifestations of the condition are present Clinically the iliac horns form bilateral symmetrical bony protuberances which are of little importance and produce no symptoms They may be recognized by palpation on physical examination This anomaly is of interest to the roentgenologist as it indicates the need for search for other clinically more significant manifestations of the syndrome They are not isolated bony malformations but comprise manifestations of the syndrome Iliac horns have been recorded in several members of the same family Anatomical studies have failed to show any connection between the exostoses and the sites for muscular attachments As a rule there are few or no symptoms referable to the defects in the bones There may be varying degrees of limitation of flexion and extension at the elbow or of pronation and supination of the forearm in patients with involvement of the elbow In the patient with defects in the patella there may be minor abnormalities of motion of the knee with instability or weakness in extension

The anomalies are considered the result of hereditary transmission of prominent characters which are not sex linked The anomaly is carried on one side from the mesoderm and on the other from the ectoderm Many genetic variations of these malformations have been described Aschner demonstrated that each of the anomalies may occur independently or in variable combinations with the other two For example a defect of the patella may occur independently without involvement of the nails or the elbow joint Hereditary dystrophy in the absence of skeletal anomalies has been noted occasionally and luxation of the head of the radius has been described as an isolated anomaly It appears that there must exist three separate pathological factors which are closely linked as the defects are often inherited together Since the entire syndrome occurs less often than the association of the defects in the nails and the patellas it must be assumed that there is a closer linkage between these two genes in order to explain their frequent occurrence together and their inheritance as if they were one factor The genes of the patella and the thumb nail are together in the same chromosome and must be very near neighbors within the chromosome The gene for the luxation of the head of the radius is probably in the neighborhood of the other two genes but farther removed in the chromosome since the whole syndrome does not occur as often as the association of the defect of the thumb nail and the patella

ADDITIONAL READING

Mino R A Mino V H and Livingstone R G Osseous Dysplasia and Dystrophy of the Nails Am J Roent *60* 633 1948

Senturia H R and Senturia B D Congenital Absence of Patellas Associated with Arthrodysplasia of Elbows and Dystrophy of the Nails Am J Roent *51* 352 1944

Thompson F A Walker E T and Weens H S Iliac Horns an Osseous Manifestation of Hereditary Arthrodysplasia Associated with Dystrophy of the Finger Nails Radiol *53* 88 1949

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MONGOLIAN IDIOCY

Mongolian idiocy is characterized by mental deficiency and a peculiar facial appearance which resembles the *mongol facies* The brain is small and the cortical pattern simplified In the past the condition has been thought to be due to defective development and to be most apt to occur in the offspring of elderly mothers, the last child commonly being affected *It is a congenital, general maldevelopment whose cause is unknown* Ingalls and Davies noted that in a group of seven cases of mongolism, in six there was a history of intercurrent maternal infections during the pregnancy which produced mongoloid children There was one case each of influenza rubella, mumps mastoiditis, pleurisy, grippe, and otitis media with sinusitis The maternal infection occurred at about the second month of pregnancy and was considered by them to be "provocative," raising the question as to whether there may not have been a correlation between the



FIG 62 Mongolism Hand wrist and forearm There is shortening of the middle phalanx of the little finger with bowing of this finger toward the ring finger which has been described as being characteristic of mongolism There is delay in the ossification of the epiphyses

maternal infection in pregnancy and mongolism However, a mother frequently gives birth to twins with but one affected indicating that infection was not the cause since it would have been expected that both twins would have been mongoloid Macklin states 'Although direct hereditary descent from parent to child is not known in mongolism, the evidence derived from a study of twins is strongly in favor of there being a genetic factor for mongolism In all instances in which twins have exhibited mongolism in one or both members of the pair monozygous twins always show both affected and dizygous twins almost always show but one affected There are a number of families on record in which several sibs have mongolism two families at least with three mongoloid children each " She concludes that mongolism which so seldom shows familial distribution is probably based on genetic factors although this does not preclude extrinsic factors from playing a role also The diagnosis is usually obvious at birth The head is small the nose flat and short the eyes slanted and the palpebral fissures narrow the entire face being flatter and smaller than

and constitutes a hereditary arthrodysplasia or familial dyschondroplasia and has been termed Turner s syndrome in the literature The bony protuberances from the iliac bones may occur as the only manifestation or in association with other changes The iliac horns are not a constant feature of the hereditary syndrome in many instances the pelvis being normal while other manifestations of the condition are present Clinically the iliac horns form bilateral symmetrical bony protuberances which are of little importance and produce no symptoms They may be recognized by palpation on physical examination This anomaly is of interest to the roentgenologist as it indicates the need for search for other clinically more significant manifestations of the syndrome They are not isolated bony malformations but comprise manifestations of the syndrome Iliac horns have been recorded in several members of the same family Anatomical studies have failed to show any connection between the exostoses and the sites for muscular attachments As a rule, there are few or no symptoms referable to the defects in the bones There may be varying degrees of limitation of flexion and extension at the elbow or of pronation and supination of the forearm in patients with involvement of the elbow In the patient with defects in the patella there may be minor abnormalities of motion of the knee with instability or weakness in extension

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ANEMIAS IN INFANCY AND CHILDHOOD

During infancy and childhood, anemia may develop insidiously and diagnosis becomes extremely difficult The problems of diagnosis are further complicated by numerous physiologic and anatomic changes within and outside of the hematopoietic system The history, the physical examination, the study of the bone marrow by sternal aspiration, the roentgen manifestations, the hereditary aspects of the disease, careful study of the blood, and the response to treatment are all important aids in arriving at a diagnosis It is essential to obtain data concerning the onset of pallor, purpura, loss of blood from the intestinal tract, infections exposure to animal parasites, the use of drugs, renal disease, rapidity of growth, overfeeding of milk with refusal of solid foods and the existence of anemia in the mother during pregnancy Information with regard to race and nationality is important in the diagnosis of sickle cell anemia and Mediterranean anemia While sickling of the red blood cells occurs almost exclusively in the Negro race, it has been observed occasionally in white families, principally those of Mediterranean origin In the Mediterranean anemias, the patients are mainly of Greek, Italian, or Syrian origin, although the disease has also occurred in the Chinese Asiatic Indian, Turkish, Egyptian and other nationalities Enlargement of the spleen is commonly found in infants and young children suffering from anemia The enlargement of the spleen is particularly prone to occur in periods of pronounced anemia and the spleen is apt to shrink during intervals of quiescence, particularly after transfusion

The value of roentgen study in the diagnosis of hematic dyscrasias in the early years of life is emphasized by the developmental features of the bone marrow It is with the appearance of nonfunctioning yellow marrow in the older child and its extension in the young adult that a potential reservoir is created for the formation of blood when the demand for hematic regeneration is increased During early infancy the need for increased blood formation arising from anemia due to infection or associated with specific hematic disease frequently necessitates the reactivation of extramedullary fetal sites With additional need for the formation of blood, the marrow expands by absorption or atrophy of bone trabeculæ and cortex and it is the demonstration of these changes on the roentgenogram which is of great importance in diagnosis Leukemia may be difficult to distinguish clinically from other hematic dyscrasias in the stages during which the anemia is associated with leukopenia and minimal enlargement of the spleen and lymph nodes Roentgen study frequently throws light on the underlying pathologic process in leukemia by the disclosure of generalized or local rarefaction, focal areas of absorption of the bone, and periosteal elevation The extensive erythrogenesis seen in Mediterranean anemia familial hemolytic jaundice and sickle-cell anemia accentuates these changes In consequence pronounced skeletal changes develop with regularity in Mediterranean anemia and similar but less pronounced manifestations are present in other members of this group In Mediterranean anemia the roentgenogram reveals evidences of extreme hyperplasia of the bone marrow with osteoporosis thinning of the cortex trabecular atrophy coarse reticulation with regeneration of new bone and thickening of the skull Regeneration of new bone is a feature of the later stages of the disease Lateral views of the skull show an enlarged diploic

usual The hair is thin and dry and the skin coarse The tongue protrudes between the lips and is thick The limbs are short, the fingers broad and stubby, and the feet are apt to be deformed Development is slow talking delayed and mental deficiency marked

Roentgen Findings The skull is small and ossification is late The sutures and fontanels may remain open until the child reaches the age of four or five years The eruption of the teeth is delayed The growth is variable, the development of the bones and epiphyses frequently being retarded and asymmetrical In other cases, however, there may be normal or accelerated growth of the bones A characteristic finding in 80 per cent of the cases is a shortening of the middle phalanx of the little finger and bowing of this phalanx toward the middle finger According to Benda the microcrania in mongolism is of the brachycephalic type

The pharynx and nasopharynx are flattened ventrodorsally There is abnormal growth and development of the skull the sphenoid being rotated upward and backward in relation to the clivus The nasion and the upper and lower maxillæ are retracted The calvaria is thin and lacks diploic structure There is delay in the pneumatization of the paranasal sinuses and late closure of the sutures



Fig 63 Sickle Cell Anemia There is granularity with multiple linear striations arranged in parallel fashion in the posterior aspect of the parietal bone producing the characteristic hair like appearance



Fig 64

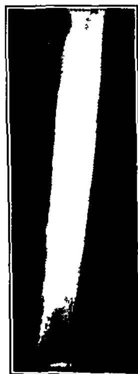


Fig 65

Fig 64 Sickle Cell Anemia The bones of the pelvis and the femurs show marked sclerosis and cortical thickening There is extensive destruction of the hip joints with aseptic necrosis of the heads of the femurs

Fig 65 Sickle Cell Anemia There is cortical thickening and irregularity of outline involving the shaft of the femur The medullary cavity is narrowed

space which is finely granular, mottled, or striated The vertical striations seem to extend beyond the outer table and give the appearance of hair standing on end The earliest manifestations are observed in the small bones, particularly the metacarpals and the metatarsals, and comprise osteoporosis and expansion of the medullary cavities producing a rectangular rather than the normal concave appearance The initial osteoporosis which occurs during infancy and early childhood is later succeeded by osteosclerosis The new bone which is formed is represented in the roentgenogram by trabeculation and coarse reticulation Osteosclerosis with cortical thickening in the large bones constitutes an outstanding feature of sickle cell anemia in adults although its evolution from a previous osteoporotic phase has been noted only infrequently

SICKLE CELL ANEMIA

Sickle cell anemia is a chronic hemolytic anemia characterized by dyspnoea pain in the abdomen and extremities ulcers of the legs epistaxis, and the presence of sickle shaped red blood cells It is a familial disease which is limited almost solely to the Negro race While approximately 7 per cent of the Negro population show the phenomenon of sickling only about 1 to 2 per cent of those with sicklemia develop the severe hemolytic disease termed sickle cell anemia There appears to be an inherited abnormality in the stroma of the red blood cells which constitutes the underlying etiologic factor of the condition the curious sickle shaped cells comprising a manifestation of the defect There is great variability in the susceptibility to the disease and its severity in the individual case Many of those affected die during infancy and childhood the terminal picture being characterized by prostration and shock with vascular collapse The disease was first described by Herrick in 1910 and increasing numbers of cases have been extensively studied and reported since that time It has apparently been in existence since ancient times the finding of characteristic alterations in the skulls of ancient Mayan and Peruvian Indians indicating that these tribes suffered with the same or a closely allied disorder

The condition has been reported as occurring at all ages from infancy to late adult life The highest incidence is in the third decade It appears to be slightly more common in females While most cases appear in the Negro race the disease has also been found in other peoples particularly those living in the Mediterranean regions The anemia varies in severity the hemoglobin in many instances being depressed to 3 0 to 8 5 grams The duration may be only a few months or as many as twenty or more years Many of those afflicted live to complete a normal life span Because of the great variability of the clinical manifestations the diagnosis is frequently not established for long periods of time one or more erroneous diagnoses being made before the correct diagnosis is achieved

Pathogenesis The cells affected in the sickling process become inflexible and fixed The length may increase to 2 to 5 times the original size and long tapering processes extend from the cells Capillary blockage results from the presence of the abnormally large cells the resultant stasis of blood in the capillaries and the small blood vessels causing thromboses and infarcts The changes in the bones are consequent upon hyperplasia of the marrow secondary to a severe degree of hemolysis congestion thrombosis, scarring and bone regeneration The nervous system may be

discs expand into the collapsing vertebral bodies and the bodies become biconcave, the so called "fish vertebra"

Sickle cell anemia is believed to be the result of a congenital anomaly of the portion of the hematopoietic system concerned with the development of erythrocytes. The abnormal cells which are produced act as foreign bodies and are destroyed prematurely by the reticulo endothelial system causing varying degrees of anemia. The bone marrow responds to overcome the anemia by an increased production of red cells, hence becomes hyperplastic and undergoes expansion. This expansion progresses at the expense of bone and produces secondary changes in the skeletal system. The bone changes occur without external enlargement except in the case of the parietal bone. There is a change in the cortical outline of the bone



Fig. 66. Sickle Cell Anemia. A Dorsal spine anteroposterior view. B Lumbar spine lateral view. The vertebræ show biconcave defects involving the superior and inferior aspects of the vertebral bodies. There is a moderate degree of generalized osteosclerosis.

indicating that the process is intraosseous. The effect of the excessive and abnormal growth activity of the marrow is most marked in the bones which are engaged in blood cell formation. In consequence, the skull and the cancellous bones of the trunk show the most extensive changes. In the segments of the long bones which usually do not participate in hematopoiesis, sclerosis predominates. This change is minimal in the distal portions of the bones of the hands and feet.

Physical Examination and Laboratory Data The patient shows stunted growth, emaciation and anemia of the oral mucosa and conjunctiva. The sclera are yellowish green, the tonsils are enlarged, and there is lymph adenopathy, particularly in the neck and axilla. The joints are swollen and painful. The heart is enlarged and there is a soft systolic murmur at the apex of the heart. The sex development may be retarded. Chronic

affected The heart is frequently involved Progressive renal damage pulmonary thrombosis and infarction, gall stones and ulcers of the skin are common The liver is enlarged and the spleen is large early in the disease becoming small in older children and adults The demonstration of the characteristic sickle cells in the red blood corpuscles establishes the diagnosis

Pathology At autopsy there are extensive vascular lesions characterized by infarction, thrombosis and focal areas of necrosis There is extensive erythrophagocytosis in the spleen, the liver the lymph nodes and the bone marrow The sickle shaped cells interlock and pack in the capillaries with resultant stasis edema and congestion This causes added sickling because of anoxemia In the bones there are degenerative processes consisting of thrombosis infarction necrosis and hemorrhage with simultaneous evidences of repair characterized by hyalinization fibrosis abnormal calcification and new bone formation The bone changes in some instances are similar to those which occur in circulatory disturbances after trauma or caisson disease Splenectomy has been advocated but does not appear to alter the course of the disease In the early phases the malpighian bodies are decreased in size fibrotic, and void of germinal centers The liver shows albuminous and fatty degeneration the sinusoids being dilated and distended with red cells There are large amounts of hemosiderin in the cells lining the sinusoids

The bone marrow is generally dark red in color and jelly like in consistency On microscopic examination there is congestion and increased cellularity with thrombosis infarction and connective tissue replacement of the parenchyma There may also be hemorrhage granular and crystalline pigment deposits hyalinization and abnormal calcifications The bone is weakened and when subjected to excessive stress shows a tendency toward compensatory new bone formation Trabecular destruction with connective tissue replacement and new bone formation in the remaining trabeculæ occur simultaneously The former produces gradual bone resorption which is manifested on the roentgenogram by a decrease in the number of bone trabeculæ and a general decrease in the density of the involved bone The latter causes an increase in the size of the remaining trabeculæ with coarsening of the trabecular structure and increase in the density of the involved bone These changes are most marked in the most erythropoietic portions of the bony skeleton that is in the cancellous bone The connective tissue replacement of the bony trabeculæ is characterized by an increase in the osteoblasts rather than by a decrease in the activity of the remaining osteoblasts In the case of the spine the coarse trabeculæ do not afford adequate strength to the osteoporotic bone particularly when it is subjected to increased stress In consequence there is a gradual collapse of the vertebral bodies This affects the dorsal and lumbar vertebræ and also in some instances the lower segments of the cervical spine The extent of the change is dependent on the duration of the disease the severity of anemia and the body weight This accounts for the occurrence of the vertebral changes in children The collapse apparently develops simultaneously in all of the affected vertebræ It may be accompanied by mild or moderately severe back pain The pain is usually transient and self limited and tends to be recurrent The collapsed vertebral bodies show increased coarseness of the trabecular structure with coarse striations which extend superiorly and inferiorly The bone density is as great or greater than that in normal bone in many instances The intervertebral

Multiple bands of increased density adjacent to the epiphyseal ends of the long bones are common and indicate periods of severe growth disturbance When roentgen manifestations occur, they indicate long standing marrow hyperplasia and infarction with osteosclerosis Partial absorption of the ungual tufts of the fingers and toes, abnormal dentition and delayed osseous union have been described

In the spine, the important alterations consist of loss of vertical height, alteration of the height-weight ratio, cupping or biconcave deformity and sclerosis of the vertebræ These changes are not seen prior to the age of thirteen They are considered specific as no other disease appears to present concomitantly hyperplasia of the marrow and multiple spotty bone infarcts which cause irregular areas of sclerosis in the vertebræ There is shortening and increase in the width of the body The changes are most marked in the lower dorsal and lumbar areas and are the result of weight bearing as the pressure of the weight of the body compresses the poorly calcified, osteoporotic vertebræ Similar changes occur in osteitis deformans, hyperparathyroidism osteomalacia due to deficiency in calcium and vitamin D, and postmenopausal osteoporosis Dorsal kyphosis develops as a sequel to the vertebral changes The teeth and jaws show generalized osteoporosis with decrease in the number of trabeculæ and increased radiolucency The remaining trabeculæ may be sharply defined and surround a multi-sized and irregularly shaped marrow space The osteoporosis affects the alveolar bone and the spongiosa of the mandible There is marked decrease in the trabeculation particularly in the bone between the roots of the teeth The trabeculæ appear to be aligned in horizontal rows, an appearance which simulates a stepladder effect In areas where no teeth are present, the trabeculæ are decreased but the parallel arrangement does not occur The lamina dura surrounding the roots of the teeth appears distinct and dense

In the chest, cardiac enlargement is the most common finding The enlargement is generalized and is independent of the patient s age or the severity of the anemia It is this manifestation in association with the murmurs which are often present which lead to an incorrect diagnosis of rheumatic heart disease in the majority of cases It is believed that the anemia causes an increase in the cardiac output with resultant dilatation and hypertrophy The osseous structures of the thorax may reveal extensive changes, there usually being generalized demineralization and coarsening of the trabecular structure While not specific in themselves the alterations in the bones together with the cardiac enlargement usually establish the diagnosis in a high percentage of the cases Splenomegaly and hepatomegaly are common and occur at all ages Splenic calcification is reported in 10 per cent of the cases The calcific depositions are amorphous and generalized The splenomegaly is less pronounced than is the enlargement of the liver and frequently the spleen becomes smaller in older children and adults Other anomalies which have been noted in association with sickle cell anemia comprise calcified biliary calculi long standing chronic osteomyelitis of the shafts of the long bones the sternum ribs etc pneumonia tuberculosis of the lungs and valvular heart disease An adult with known severe anemia for many years may show no alterations on roentgen study while younger patients with clinically milder disease present marked osseous changes The severity of the anemia or the frequency of the crises is not the determining factor although the chronicity does appear important in this regard

ulcers of the legs, with a punched out appearance which closely resembles syphilis, are an almost constant finding above the age of sixteen The red cell count is diminished to about 3,000 000 and in some instances to less than one million The hemoglobin is reduced in very severe cases and may be as low as 10 per cent The color index is usually normal There is moderate leukocytosis in some cases with increase in the neutrophiles In the presence of intercurrent infection the white cell count may be elevated to 50,000 or more The blood smear shows sickling of the red cells These sickle cells are best seen in the twenty four hour preparation The icterus index is increased, as are also the phosphatase and blood calcium in the active stages of the disease The bleeding time and the coagulation time are normal

Prognosis The prognosis is very poor in severe cases in infancy and childhood In the milder cases the lower resistance to intercurrent infections usually results fatally and the patients rarely live beyond the age of thirty

Roentgen Manifestations Roentgen examination is of great importance in sickle cell anemia as the extent of the manifestations roughly parallels and can be correlated with the duration and severity of the disease The skull is the most frequently involved of the bones The earliest change is the development of a ground glass appearance The most characteristic findings comprise a marked cortical thickening and spicules of bone radiating from the bones producing a hair on end appearance The changes occur within and beyond the thickened outer table during childhood after the fifth year of life and are due to the presence of trabecular striations which radiate outward perpendicular to the inner table Later in life, the striæ are less frequently seen lamellated new bone formation being present This manifestation is relatively uncommon and is not entirely pathognomonic as it occurs also in chronic hemolytic icterus, erythroblastic anemia rickets and scurvy The biparietal diameter of the skull is increased due to an actual increase in the width of the parietal bones The changes in the skull vary with the stage of the disease as well as the age of the patient The bones may be granular and mottled The diploic spaces are widened and dense and/or may be obliterated The differentiation between the inner and outer tables is lost An oxycephalic appearance or tower skull may be present

In the long bones during childhood there occur coarsening of the trabeculæ widening of the medullary spaces and cortical thinning The spicules which remain in the marrow cavity become more conspicuous In adults, the roentgen examination may be non contributory no changes being present in many instances In other cases there occur narrowing of the medullary cavity loss of bone tissue periosteal reaction and cortical thickening in the long bones This probably is due to the presence of multiple thromboses The cortical thickening may be patchy with scattered small areas of increased radiance Joint destruction may ensue if the thrombosis interferes with the blood supply to the joint Infarction of the femoral head produces osteoarthritic changes aseptic necrosis and changes similar to those in Legg Perthes disease The proximal epiphysis of the humerus and other bones may be similarly affected Long standing infection and vascular stasis in the cutaneous and subcutaneous tissues cause periosteal changes in the underlying bone The chronic leg ulcers of sickle cell anemia may also be associated with gross roughening and irregularity of outline of the adjacent bone due to periosteal proliferation

On roentgen study, the disease is characterized by diffuse bone changes The medullary portions of the bones are widened and the trabeculations are increased, particularly at the ends of the diaphyses The cortex is markedly thinned and pathologic fractures are common Periosteal elevation is rare Joint changes may occur The skull in early cases shows only slight thickening of the vault and osteoporosis As the disease ad

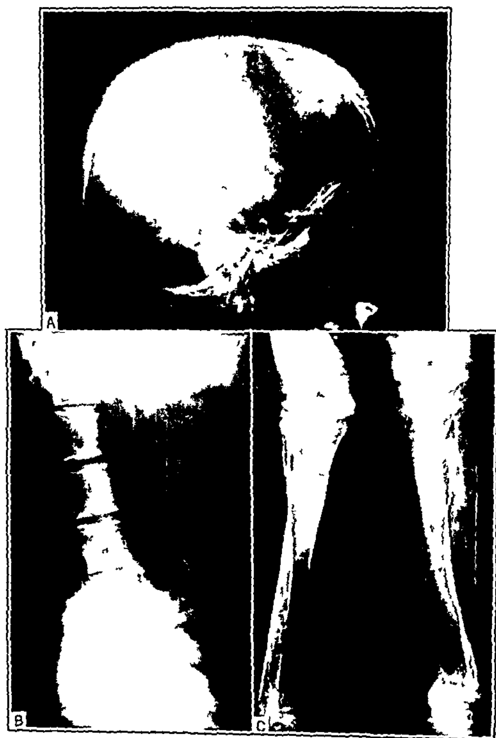


Fig. 67 Cooley's (Erythroblastic) Anemia *A* Skull lateral view There is thickening of the inner table with linear striations in the parietal region *B* Lumbar spine lateral view The vertebræ show generalized osteoporosis The intervertebral spaces are preserved and there is no collapse of the vertebræ *C* Lower legs The medullary portions of the bones are markedly widened and the cortex is thinned There is marked osteoporosis The trabeculæ are obliterated There are transverse metaphyseal lines

Differential Diagnosis Sickle cell anemia cannot be differentiated from caisson disease by roentgen study alone While it is not possible to establish the diagnosis with definiteness by roentgen methods the disease should definitely be suspected despite the fact that similar bone changes are found in any anemia of congenital origin The skull changes are similar to those in erythroblastic anemia or congenital hemolytic icterus In sickle cell anemia the parietal bones show more extensive involvement than the frontals while the reverse is true in other congenital anemias The bones of the hands and feet are affected more often in sickle cell anemia In later life, because of the osteosclerosis, the differential diagnosis must include osteoblastic metastatic disease atypical hyperparathyroidism, osteitis deformans, polyostotic fibrous dysplasia marble bone disease, fluorine intoxication leukemia, and aplastic anemia It is also necessary to consider syphilis tuberculosis and Legg Perthes disease The changes in the spine are similar to those in senile osteoporosis the collapse being preceded by generalized bone atrophy and decalcification It is not associated with coarsening of the trabeculae In senile collapse there are often pathological compression fractures This has not been noted in sickle cell anemia Similar changes do not occur in the vertebral bodies in Cooley's anemia or congenital hemolytic anemia This is surprising as the underlying pathology is similar in the three diseases There is definite similarity of the osseous manifestations in sickle cell anemia and Paget's disease There is increase of the serum phosphatase level The bone marrow is hyperemic in both In Paget's disease the serum calcium and inorganic phosphorus levels are practically always normal The alkaline serum phosphatase is elevated in proportion to the extent of osteoblastic bone involvement In osteoporosis circumscripta an early stage of Paget's disease there is bone destruction The serum phosphatase is normal and there is decreased function of the osteoblasts in the involved area Roentgen examination of the lumbar vertebræ often shows vertebral flattening very similar to that which occurs in sickle cell anemia except that the structure of the trabeculae is coarser

ERYTHROBLASTIC ANEMIA COOLEY'S ANEMIA

Erythroblastic anemia is usually seen in infants and children of Greek and Italian parentage The condition is congenital and familial The etiology is unknown There is enlargement of the parietal portions of the skull and the features are mongoloid The disease is usually well established in the first year of life and the mortality is high It is due to a disturbance of the hematopoietic system with progressive anemia and is characterized by large numbers of nucleated red blood cells in the blood stream Leukocytosis hepatomegaly and splenomegaly occur There are large numbers of erythroblasts and many immature red cells are present There is a mild degree of icterus, probably the result of hemolysis The color of the scleræ remains normal The bones are extremely thin and the red marrow can be seen on elevation or removal of the periosteum The marrow cavity is widened and filled with dense red tissue The inner and outer tables of the skull are very thin and there is marked porosity of the medulla Early the bones appear mottled later they are markedly thickened

increased to a degree which produces significant ocular hypertelorism The lesions in the tubular bones of the extremities regress with increase of age while the changes in the central segments of the skeleton persist and increase The regression first becomes evident at the onset of pubescence The pneumatization of the paranasal sinuses and the temporal bone appears to be retarded in all cases After puberty, the central portions of the skeleton, that is the skull, spine and pelvic bones, are the optimal sites for demonstration of roentgen changes in the bones The bones of the hand, which are the most important sites for roentgen demonstration of Cooley's anemia in children, become the poorest sites after puberty

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ERYTHROBLASTOSIS FETALIS

Erythroblastosis fetalis is characterized by generalized edema, jaundice, anemia and erythroblastemia Intra uterine death with maceration of the fetus may occur as a manifestation of this disease The fetus may show enlargement of the liver and spleen, extramedullary erythropoiesis, and changes in the bones The condition usually occurs in children born of mothers who are Rh negative and fathers who are Rh positive While Rh negative women mate with Rh positive men in approximately 10 per cent of all marriages congenital hemolytic disease appears to occur in approximately only 1 of 250 newborns Although there is no method of combating certain fetal complications of the disease many infants with erythroblastosis fetalis may be saved by transfusions of Rh negative blood other than that of the mother or washed maternal cells Early recognition of the condition is of the utmost importance for the prompt institution of effective therapy Roentgen examination of the fetus *in utero* affords valuable information and may permit of a definite diagnosis in certain instances

Erythroblastosis fetalis occurs in three forms (1) fetal hydrops, (2) icterus gravis, and (3) congenital anemia (1) The first is characterized by accumulation of fluid in the soft tissues and body cavities enlargement of the liver and spleen anemia immature red cells, and a large placenta This type is very severe and is usually fatal during the intrauterine period or within a few hours after birth (2) The second form, icterus gravis, is usually associated with jaundice at the time of birth or within the first twenty-four hours, although milder forms occur in which this manifestation does not develop until later in some instances only after one to two weeks Usually the jaundice increases in intensity and there is progressive anemia Pulmonary hemorrhage or injury to the brain results in death In the less severe cases the jaundice fades in one to two weeks (3) Congenital anemia is the mildest and least frequent type of erythroblastosis fetalis The only sign in this form of the disease is extreme pallor

The Rh factor is a mendelian dominant Hence, an Rh negative woman married to an Rh positive man may expect all the offspring to be Rh posi-

vances, both the inner and outer tables show marked thinning with simultaneously marked increase in the thickness and density of the medullary portions. The inner table may remain dense, while the outer table in some instances is so thin as to be scarcely visible. The diploe is markedly thickened, being increased to several times its normal thickness. No periosteal proliferation occurs. The skull in the late stages of the disease shows thickening of the bones especially in the parietal region. The thickening involves the diploe, the diploic spaces being obliterated and the inner and outer tables becoming indistinguishable. The increased density is homogeneous. Vertical striations occur in the skull with bony spicules radiating at right angles to the outer table. The thin, hair like spicules radiating perpendicularly to the bones of the skull have the appearance of hair standing erect and are most marked in the parietal region, the frontal and temporal bones being less frequently involved. The change is very similar to that in sickle cell anemia.

Caffey reports a series of observations on four patients with Cooley's anemia who were studied during prolonged periods from early childhood into the second and third decades of life. In the distal portions of the skeleton, that is the hands, the arms and the legs, the bone lesions tended to regress during late childhood. However, during the same period, similar changes persisted and apparently showed increase in the calvaria, the pelvis, the long bones, and the central portions of the skeleton. The skeletal changes in Cooley's anemia appear to be due to overactivity and overgrowth of the bone marrow. The regression distally and persistence centrally follows the pattern of regression of the normal marrow from the distal portions of the skeleton toward the trunk with advancing age. In some instances the regression becomes evident during pubescence. This process does not proceed uniformly in all individuals, in some cases being slower and less complete than in others. It may be altered by blood transfusions. The earlier activity of the pubescent hormonal factors in females may be responsible for differences which occur in the two sexes. Regressions occur in the ribs but these bones are not affected as early or as markedly as the tubular bones of the extremities.

Pneumatization of the cranial air sinuses may be affected. Inhibition of pneumatization may become evident as early as the fourth year and the retardation of the pneumatization becomes more pronounced as the age of the patient increases. The maxillary and sphenoidal sinuses are small in every instance and in some cases are completely obliterated. The frontal sinuses are small but are not as markedly retarded as the maxillary and sphenoidal sinuses. The ethmoidal cells appear normal, indicative of the fact that there is little or no marrow activity in the ethmoid bone. Pneumatization of the temporal bone is almost completely absent. This is manifested by absence of the air cells in the mastoids and the zygomatic and squamous segments. The petrous pyramids may also show absence of air cells. There is a disproportionate overgrowth of the upper maxilla with ventral and lateral expansion of the alveolar ridges and resultant malocclusion of the jaws. Other manifestations comprise ventral protrusion of the upper central incisor teeth with retraction of the upper lips and overbite on the lower jaw giving rise to a rodent facies in older children. This is more common and more diagnostic of Cooley's anemia than the mongoloid facies which is characteristic of the disease in certain younger children. Because of the marked swelling of the upper maxilla the orbit is often shifted laterally and the intraorbital distance

be obscured *in utero* by the amniotic fluid and the overlying densities of the maternal structures

(2) *Skeletal Abnormalities* The osseous changes in erythroblastosis fetalis were first described by Follis, Jackson and Carnes and consist of a marked increase in the density of the bones The skull, vertebræ, ribs, pelvis and long bones may be affected in varying degrees In the skull the sphenoid and occiput are principally involved, the remainder of the calvaria showing less or no involvement The vertebræ show marked sclerosis In the ribs, the density is most clearly visualized at the angles The pelvis and other flat bones may be diffusely eburnated The long



Fig 70



Fig 71

Fig 70 Osteosclerosis in Erythroblastosis Fetali The bones of the legs are increased in density The changes are generalized and uniform

Fig 71 Erythroblastosis Fetalis The bones of the base the occipital bone the facial bones and the cervical vertebræ are extremely dense There is overlapping of the bones of the skull There is very marked edema of the scalp and chin

bones in the cases observed by us revealed uniformly increased density throughout the entire shafts with marked narrowing or obliteration of the medullary spaces Follis Jackson and Carnes described zones of diminished density of varying width at the ends of the tubular bones immedi ately below the cartilage shaft junctions On carefully executed roent genograms the abnormalities in the bones may be demonstrable *in utero* This may be of great value in diagnosis as it enables the clinician to determine during the course of the pregnancy whether the fetus is erythroblastic or normal The change in the density of the bones is demonstrable both on the roentgenogram and microscopically There is an actual increase in the number and thickness of the bone trabeculæ This is ap

tive if the husband is homozygous for Rh, and half to be Rh positive if the husband is heterozygous for Rh Reactions involving anti-Rh agglutinins rarely occur unless the woman has been prgenant or has been transfused previously However once she has developed a concentration of antibodies sufficient to produce erythroblastosis, all subsequent offspring will be affected The occurrence of hemolytic disease varies widely in different series which have been reported ranging from two to five per cent of the children born to Rh negative mothers This indicates that Rh iso immunization is not in every instance associated with hemolytic disease of the



FIG 68 FIG 69

FIG 68 Erythroblastosis Fetalis Diagnosed *in utero* Eleven days prior to delivery There is extensive sclerosis involving practically the entire skeleton

FIG 69 Erythroblastosis Fetalis The bones of the pelvis the vertebræ the ribs clavicles scapulæ and the long bones show increased density cortical thickening and narrowing of the medullary spaces The liver and spleen are markedly enlarged

newborn The male fetus is apparently more susceptible to erythroblastosis fetalis than the female Twins may not both be affected in some instances one being Rh positive and the other Rh negative

The roentgen findings in the cases of erythroblastosis fetalis may be divided into three main groups (1) soft tissue changes consisting of generalized edema and enlargement of the liver and spleen (2) abnormalities of the skeleton and (3) evidences of fetal death

(1) *Soft Tissue Changes* There occurs massive edema with very marked swelling and thickening of the soft tissues of the head face and neck The edematous fleshy tissues of the body and limbs appear thicker and denser than normal on the roentgenogram The changes in the soft tissues may

ess and ossified stylohyoid ligaments Unusually long styloid processes are encountered in approximately one out of every three thousand tonsilec tomies Forty-four cases of complete ossification of the stylohyoid ligaments have been recorded In man the hyoid bone is the only bone which normally forms no joint with other bones of the body It has a considerable range of upward, downward, forward, and backward mo bility If the stylohyoid or the hyothyroid ligaments become extensively ossified, the mobility of the hyoid bone is interfered with, the hyoid becomes less flexible and unusual stresses in this region may produce paresthesias in the neck or throat with a sense of fullness, burning or pressure There may also be pains in the ears, neck, throat, tonsils and back of the tongue with radiation to the external occipital protuberance Other mani festations comprise persistent cough or paroxysms of coughing, dysphagia, and dysphonia In case of penetration or perforation of the stylohyoid processes the symptoms may persist for weeks or months, although usually the duration is but a few days Elongation of the styloid processes may cause the processes to protrude from the tonsils, the lateral wall of the pharynx, or the base of the tongue If the process is not visible, palpation may disclose a bony resistance and be attended by pain radiating to the area of the external occipital protuberance Roentgenography and fluoroscopy are important aids in studying the hyoid bone and should be carried out in all cases of unexplained dysphagia An important feature is the long clinical course, the symptoms usually persisting for many years With markedly elongated styloid processes or ossified stylohyoid ligaments, stresses such as a sudden twist of the head produce pain

ANOMALIES OF THE RIBS

Anomalies of the ribs are relatively common It is important that the roentgenologist be familiar with the various rib anomalies and distinguish those which are developmental in origin from others associated with functional defects or disease Anomalies of the first rib appear to occur in about 0 15 per cent to 1 0 per cent of all individuals Asymmetry of development is the most frequent normal deviation Another common finding is failure of ossification of the anterior portion of the first rib, usually in the region anterior to the scalene tubercle Todd states that this occurs during embryological development and is due to compression of the subclavian muscles in the grooves in the upper aspect of the first rib Ribs so deformed are short and may simulate cervical ribs In order to establish the diagnosis it may be necessary to examine the entire cervical dorsal and lumbar spine Partial ossification of the anterior portions of the first ribs is most apt to occur in the region of the manubrium with resultant deficiency of the sternoclavicular joint The presence of an incomplete foramen indicative of a pre-bifid anomaly is not infrequent and affects the ribs in the region of the costochondral junction Complete absence of the first rib has been described There is no explanation of the cause of this defect Fusion of part or all of the first rib with the second rib is not infrequent A theory to explain these rib mutations has been to link them with evolution of disease in the underlying lung or pleura particularly tuberculosis However, careful study of large series of cases has not shown any association between tuberculosis or other diseases in the upper lobes and anomalies of the first ribs

parently due to lack of destruction of the calcified cartilaginous matrix substance which is then covered with a thick layer of bone In congenital lues there is increased density of the bones Also in osteopetrosis (Albers Schonberg disease), known as osteosclerosis fragilis generalisata there are bone changes very similar to those in erythroblastosis fetalis However multiple fractures are common in osteopetrosis but have not been noted in erythroblastosis

(3) *Evidences of Fetal Death* Death of the fetus occurs with resultant stillbirth in many cases of erythroblastosis fetalis Overlapping of the bones of the skull is the earliest and most reliable sign that the fetus has died During active labor, the bones of the fetal skull may undergo varying degrees of compression and overlap slightly at the sutures Except when the patient is in labor however overlapping of the skull bones is considered pathognomonic of fetal death This change may occur within a few days or weeks after the cessation of life Sharp angulation of the head in relation to the spine, lordosis of the caudal extremity of the spinal column collapse of the thoracic cage and marked, generalized decalcification of the fetal skeleton are also accepted as roentgen manifestations of death of the fetus these are later in developing and are not always dependable criteria however Marked disproportion between the size of the fetus and its expected development at the supposed clinical duration of the pregnancy is significant in this respect

The demonstration of the above described changes *in utero* is difficult The utmost care is essential in the production of as nearly technically perfect roentgenograms as possible High milliamperage technique is necessary A short exposure is essential to eliminate the possibility of fetal movement during the making of the roentgenogram The patient must be cooperative and suspend movement and respiration completely during the roentgen exposure Anteroposterior oblique and lateral projections are made of each patient The edema of the fetal tissues and the hydramnios which are so often present in this condition are very apt to obscure the alterations in the densities of the bones and soft tissues of the fetus or the roentgen manifestations of fetal death The first roentgenograms should be made approximately ten to fourteen weeks prior to term and thereafter at intervals of three to six weeks A sudden change in titer clinical evidence of fetal death or signs of toxemia are important indications for careful roentgen studies of the fetus As with other rare and unusual conditions the diagnosis will be made only if the manifestations are clearly understood constantly borne in mind and painstakingly sought for in every instance The importance of early diagnosis in the planning of therapy and making a proper prognosis is self evident

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ANOMALIES OF THE HYOID BONE

Anomalies of the hyoid bone are extremely rare Most reports in the medical literature deal almost entirely with elongation of the styloid proc

ess and ossified stylohyoid ligaments Unusually long styloid processes are encountered in approximately one out of every three thousand tonsillectomies Forty four cases of complete ossification of the stylohyoid ligaments have been recorded In man the hyoid bone is the only bone which normally forms no joint with other bones of the body It has a considerable range of upward, downward, forward, and backward mobility If the stylohyoid or the hyothyroid ligaments become extensively ossified, the mobility of the hyoid bone is interfered with, the hyoid becomes less flexible, and unusual stresses in this region may produce paresthesias in the neck or throat with a sense of fullness, burning or pressure There may also be pains in the ears, neck, throat, tonsils and back of the tongue with radiation to the external occipital protuberance Other manifestations comprise persistent cough or paroxysms of coughing, dysphagia, and dysphonia In case of penetration or perforation of the stylohyoid processes the symptoms may persist for weeks or months, although usually the duration is but a few days Elongation of the styloid processes may cause the processes to protrude from the tonsils, the lateral wall of the pharynx, or the base of the tongue If the process is not visible, palpation may disclose a bony resistance and be attended by pain radiating to the area of the external occipital protuberance Roentgenography and fluoroscopy are important aids in studying the hyoid bone and should be carried out in all cases of unexplained dysphagia An important feature is the long clinical course, the symptoms usually persisting for many years With markedly elongated styloid processes or ossified stylohyoid ligaments, stresses such as a sudden twist of the head produce pain

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A rare anomaly of development of the first rib is pseudarthrosis This may involve cervical ribs and the first true ribs and is difficult to differentiate from a simple or pathologic fracture The pseudarthroses tend to occur in the posterior third of the first rib It has been stated that they represent fractures due to muscle traction with subsequent pseudarthrosis or a congenital anomaly similar to the costal articulation in avians The absence of callus and the smooth contours usually establish the diagnosis Roentgen examination may reveal a pseudarthrosis of the first rib in patients who give no history of injury and have no symptoms referable to this area

Synostosis of the Ribs

Synostoses of the ribs occur more commonly on the right than on the left and usually involve the first and second ribs The synostosis may be both anterior and posterior and may involve all the ribs visible on the

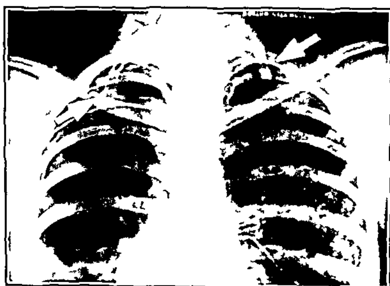


Fig 72 Cervical Ribs There are bilateral cervical ribs *The supernumerary ribs are long and well formed (arrows)*

chest roentgenogram In rare instances there may be multiple synostoses in addition to bilateral cervical ribs Occasionally the synostosis has the appearance of a joint between the ribs The synostosis may give rise to an appearance which is closely similar to that of a cyst or an emphysematous bulla Bifid ribs have the appearance of a lobster claw and are occasionally incomplete There also may be simple forking and spatula forms These occur more frequently on the right than on the left the right 4th rib being the most frequently involved Bilateral bifid ribs are rare Absence of a rib or the greater portion of a rib is not infrequent as the result of thoracic surgery or the transthoracic approach to a lesion in the abdomen After resection of a segment of a rib a synostosis may develop between the resected rib and those immediately adjacent to it

Supernumerary Ribs

Cervical ribs are a frequent cause of pain which may radiate to the shoulder arm or fingers paresthesia atrophy of the muscles of the arm

and hand, and cyanosis The condition may be unilateral or bilateral and is less common in males Symptoms often do not occur until the middle or later years of life Physical examination in some instances reveals a fullness or slight pulsation in the region of the base of the neck, although more commonly there are no clinical findings Roentgen examination is essen-

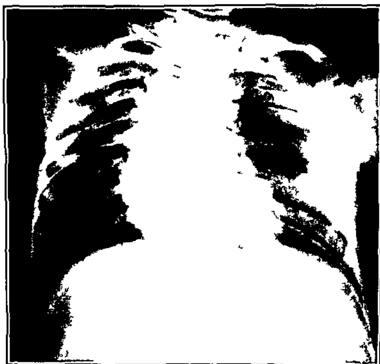


FIG 73 Congenital Anomaly of the Ribs and Spine There are multiple congenital malformations of the ribs There is a marked degree of scoliosis and deformity of the chest

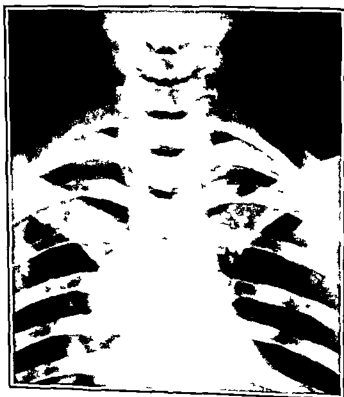


FIG 74 Bilateral Cervical Ribs

tial to the demonstration of cervical ribs When supernumerary ribs are suspected it is necessary to carry out x-ray studies of the entire thorax with films which outline the ribs and spine with a sufficient degree of clarity to permit of accurate observation of the bony structures They are frequently found on routine chest and spine films Cervical ribs may be small or large, in some instances extending anteriorly to the manubrium sterni In many cases the supernumerary rib has the width and contours of a normal rib, varying only as regards its length, frequently, however, it is narrower, less dense, and irregular in outline The supernumerary rib may fuse with the first true rib In rare cases small portions of the cervical rib may be present only at the vertebral and sternal regions When bilateral, one side is frequently larger than the other The transverse process of the seventh cervical vertebra lacks a vertebral foramen and closely resembles a dorsal vertebra in the presence of an anomalous rib The finding of a supernumerary rib is a simple but important roentgen diagnosis The condition is frequently overlooked, particularly in poorly positioned or underexposed films The importance of correct diagnosis can not be over estimated, as many unexplained symptoms may be clarified by the demonstration of this anomaly Two ribs may arise at a single costovertebral junction or a single rib may bifurcate at or near its anterior end in the costochondral region The adjacent rib may be absent or underdeveloped

Certain criteria must be present to establish the diagnosis of cervical ribs The rib must arise from the transverse process of the seventh cervical vertebra This process projects horizontally from the spine in contradistinction to the transverse process of the first dorsal vertebra which extends diagonally upward from the point of origin The rib must have no connection with the manubrium of the sternum anteriorly although it may form a synostosis with the first rib Many ribs which appear to be supernumerary are found to be rudimentary first ribs when the above criteria are applied All patients with shoulder pain should have roentgen studies of the cervical spine as well as the shoulder girdle If it reaches far enough forward part of the brachial plexus and the subclavian artery and vein cross over the cervical rib Pressure on the artery may obstruct the circulation so much that arterial thrombosis results causing gangrene of the finger tips Pressure on the nerves is commoner and affects the eighth cervical and first thoracic nerves causing paralysis of the muscles they supply and neuralgic pains and paresthesia in the area of skin to which they are distributed no oculopupillary changes are to be found Little was known about cervical ribs prior to the advent of roentgenography Identification of these anomalies by dissection is frequently impossible This accounts for the lack of data in the anatomical literature and text books prior to the widespread use of the roentgen method of diagnosis

In the lumbar region supernumerary ribs are an interesting anomaly but of no clinical significance except in so far as they may be confused with fractures or exostoses as they cause no pain or other symptoms

Rib Notching

For many years it was generally accepted that notching of the ribs occurred only in coarctation of the aorta and was pathognomonic of this condition While coarctation is the most common cause it is now known that other conditions may produce a similar change Rossler first de

scribed notching in coarctation of the aorta in 1923 and numerous cases have been recorded since that time It has been shown that the notching is present in about three fourths of the cases Dussaillant *et al* in 1945 described a case characterized by notching of the ribs in the absence of coarctation in a person with arteriosclerosis, hypertension, and emphysema Barden and Batchelder and Williams have recorded instances of unilateral rib notching associated with tortuous intercostal arteries in patients with the tetralogy of Fallot Von Recklinghausen s neurofibromatosis may be associated with changes in the ribs similar to the vascular erosions which occur in coarctation of the aorta In coarctation of the aorta, the erosions are ascribed to the abnormally large intercostal arteries pulsating in close relation to the inferior margins of the ribs The typical



FIG 75 Notching of the Ribs Coarctation of the Aorta The roentgen diagnosis of coarctation of the aorta can be made with definitene s by the demonstration of notching along the inferior margin of the ribs The fourth to eight right ribs and the fifth to ninth left ribs show narrowing irregularity of outline and notching along the inferior margins (white arrows) The aorta is definitely mall and the supracardiac area is decreased in width The heart shadow is not enlarged

case presents notching involving the third to tenth ribs bilaterally The changes are widely variable in character and extent being dependent on the degree of the coarctation its site and other factors With *constriction* of the aorta proximal to the origin of the left subclavian artery, unilateral notching involving only the right side is the rule Arterial injection studies by Bramwell and Jones demonstrated that the increased flow of blood in the tortuous dilated intercostal arteries is the primary factor in the causation of the notching Slight degrees of dilatation of the arteries may produce no alterations in the ribs The varying degrees of tortuosity and dilatation of the arteries result in unpredictable patterns of notching

In some cases of tetralogy of Fallot, there occurs an anastomosis of the intercostal arteries and the mediastinal arteries with resultant by passing of the pulmonic stenosis This in association with the bronchial arteries

forms a collateral circulation The resultant dilatation and tortuosity of the intercostal arteries may produce varying degrees of rib notching A localized tortuosity and dilatation of the intercostal arteries with rib notching may occur with no other demonstrable pathology McCord and Bavendam report a case of rib notching caused by dilated tortuous venous channels They are of the opinion that rib notching in patients without coarctation of the aorta may be associated with arteriosclerosis and a wide pulse pressure due to hypertension or aortic insufficiency resulting in unusually forceful pulsations of tortuous intercostal arteries against the ribs There are on record numerous instances in which no cause of rib notching can be determined It may occur in cases of aortic valvular disease with or without hypertension A unique case has been reported in which notching was caused by dilated tortuous intercostal veins in a patient with long standing obstruction of the superior vena cava with extensive collateral circulation It is important to stress that notching of the ribs is not pathognomonic of coarctation of the aorta or of dilatation of the intercostal arteries Angiography is essential to determine whether the vascular dilatation and tortuosity are arterial or venous in origin

CONGENITAL ELEVATION OF THE SCAPULA (SPRENGEL S DEFORMITY)

Congenital elevation of the scapula is known as Sprengel s deformity The condition is characterized by the fact that the scapula is elevated adducted and rotated downward In most instances it occurs in association with other anomalies such as malformations of the ribs anomalies of the vertebral column and asymmetry of the chest walls Lateral curvature of the spine frequently occurs and may be associated with hemivertebrae or incomplete segmentation of the vertebral bodies An omovertebral bone is frequently present and appears as a roughly trapezoid or wedge shaped structure composed of bone and cartilage The narrow extremity of this bone is continuous with a spinous process lamina or transverse process of one or more of the lower cervical vertebrae The omovertebral bone is frequently attached to the medial border of the scapula the connection being bony or cartilaginous Motion of the shoulder in patients with congenital elevation of the scapula may be only slightly impaired The deformity is usually a permanent one and does not undergo spontaneous improvement The roentgen manifestations are characteristic and permit of a definite diagnosis The affected scapula is markedly decreased in size elevated and rotated the angle of the scapula usually being directed medialward The glenoid may be small and shallow The acromioclavicular articulation lies at a higher level than normally Associated anomalies of the ribs and spine are frequent Scoliosis of the upper dorsal spine and hemivertebrae in the lower cervical and upper dorsal regions appear in a high percentage of the cases The chest is asymmetrical the affected side being decreased in size Torticollis is present in many instances Congenital elevation of the scapula is a relatively uncommon condition It appears to occur more commonly in females than in males The deformity has been noted with equal frequency on the left and right sides In rare instances it may be present bilaterally Correction of the undescended scapula can be accomplished by surgical operation The result is not good in many instances The best results are obtained in patients in whom the scapula is transplanted and replaced into its normal position early in life particularly before the age of five years

Fig 76



Fig 77

Fig 76 Sprengel s Deformity with Omovertebral Bone The left scapula is elevated adducted and rotated it is markedly reduced in size The left glenoid is small and shallow The omovertebral bone is visualized as an area of calcific density in the region adjacent to the superomedial border of the anomalous scapula There is scoliosis and spina bifida involving the upper dorsal vertebrae

Fig 77 Sprengel s Deformity The left scapula is elevated and rotated its angle being directed toward the spine The left glenoid is small and the acromioclavicular joint is elevated The chest is asymmetrical and the left upper ribs are flattened

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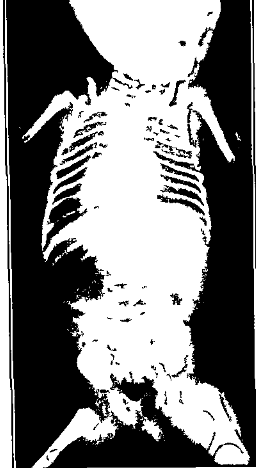


FIG 78 Congenital Anomalies Absence of the Wrists and Hands The child was born with short maldeveloped arms The hands and wrists are completely absent In each forearm a solitary rudimentary bone is present The elbow joints are not developed The remainder of the skeleton revealed no demonstrable abnormality



FIG 79 Congenital Anomaly of the Arm Ulnar Dorsal Talipomanus

CONGENITAL DEFECTS OF THE ARM

Congenital absence of the radius is relatively common and many cases have been recorded in the literature. The condition is generally associated with club hand. Absence of the radius is believed to occur more frequently than absence of the ulna and total absence is more common than partial absence. In about 50 per cent of the cases the defect is bilateral. Although any part of the radius may be missing the presence of a remnant of the upper end is frequent. The radial bones of the carpus may be absent or fused. The soft parts of the radial side of the forearm may be affected. The ulna may be shortened, thickened, and curved. As the patient grows, the hand may become deviated radially until it lies at a right angle to the long axis of the forearm. Treatment consists of release of contractures and arthrodesis in the position of function. Albee has recommended the use of a tibial graft mortised into the carpus and the shaft of the ulna to form a strut. Correction of bowing of the ulna can be performed by an osteotomy. In a case described by Barsky, a two year-old child presented an upper extremity measuring 9½ inches in length. The ulna was absent and the radius was fused with the humerus. There were but two fingers on each upper extremity and two metacarpals. The fingers contained two phalanges and were webbed. In addition to the defects in the upper extremity the patient had congenital dislocation of the left hip and cryptorchidism. There have been cases reported in which the arm or the leg was absent but a hand or foot was present. This condition is termed phocomelus.

MADELUNG'S DEFORMITY

Madelung's deformity is characterized by volar flexion and subluxation of the hand at the wrist joint. The styloid process of the ulna is prominent dorsally. The radius is usually curved, the bowing being dorsolaterally. The patient is unable to abduct the hand in the radial direction. Madelung considered the condition a spontaneous volar subluxation of the hand. However, the underlying factor comprises a growth disturbance of the lower end of the radius, the malposition of the hand being secondary to the anomaly of the radius. Other congenital malformations may also occur, particularly in the hand. It is frequently bilateral.

Madelung's deformity is divided into two groups, the true form and the symptomatic variety. The former is hereditary. The symptomatic type is due to fracture or disease of the lower end of the radius. Hereditary or familial factors are present in about 40 per cent of the published cases. The occurrence of the anomaly in two or three generations is not uncommon and the lesion has been described in six generations of one family. It appears to be a dominant, non sex linked hereditary factor. The condition constitutes reduction of growth of the distal epiphyseal disc of the radius, the volar ulnar segment particularly being affected. Premature fusion of this portion of the bone is not the cause as the condition may occur even in the presence of a completely open disc. There is angular deformity of the proximal row of carpal bones secondary to the changes in the radius.

Roentgen examination shows sharp angulation of the lower ends of the radius and ulna toward each other. The radius is shortened, bowed and

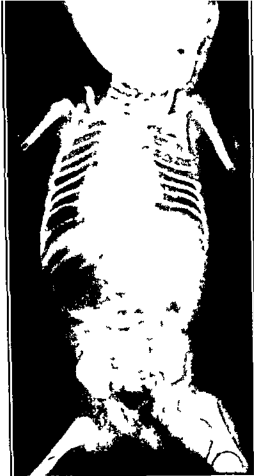


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SUPERNUMERARY CARPAL BONES

A variety of supernumerary bones may occur in the upper and lower extremities The partite navicular and partite lunate are the most common and will be discussed in detail subsequently The other supernumerary bones which may be present in the region of the wrist are relatively rare and comprise the following

external radius, at the tip of the styloid of the radius
central bone distal aspect of the navicular,
epipyramis distal portion of the lunate ulnar aspect,
partite triangular
lesser triangular, between the ulna and the lunate
intermediate antebrachial (triangulare) proximal aspect of the triangular bone
external ulna ulnar aspect of the triangular bone
secondary pisiform adjacent to the tip of the ulnar styloid
epitrapezium proximal aspect of the greater multangular,
paratrapezium radial aspect of the greater multangular
secondary greater multangular distal portion of the greater multangular ulnar aspect,
partite (bipartite) lesser multangular
secondary lesser multangular distal portion of the lesser multangular,
secondary capitate distal portion of the capitate ulnar aspect,
subcapitate distal aspect of the capitate radial aspect,
styloid distal portion of the capitate ulnar aspect
parastyloid distal portion of the greater multangular ulnar aspect
metastyloid mid portion of the capitate radial aspect
Gruber s bone distal aspect of the capitate ulnar side
subcapitate distal end of the capitate
Vesalius at the base of the fifth metacarpal ulnar aspect
basal hamular distal aspect of the hamate
hamulate distal aspect of the hamate ulnar side

Partite Navicular, Bipartite Navicular, Tripartite Navicular

Developmental variations of the human carpus are unusual The most frequent are the bipartate navicular the *os centrale* and fusions of two or more carpal bones In many instances these anatomical variations are bilateral The bipartite carpal navicular is of particular interest because the condition is easily mistaken for an ununited fracture The error stems from the fact that the line of division between the two segments of a bipartite navicular is usually at the waist of the bone which is frequently the site of an ununited fracture In many instances there is failure to obtain a roentgenogram of the opposite wrist, hence it is not realized that the anomaly is bilateral The condition has been estimated as occurring only about once in 700 cases Some authors have stated that bipartite carpal navicular represents an old unrecognized fracture followed by pseudarthrosis rather than a true anomaly of the bone However the condition is a true anomaly While partition of the navicular does occur as a developmental reversion it is probably true that many of the cases previously described have been overlooked fractures

increased in density The distal ends of the radius and ulna are incompletely formed, small in size, and may be separated from each other and dislocated from the bones of the wrist The lunate bone may be displaced toward the radial aspect of the wrist and the proximal row of carpals are compressed into a smaller space than normally and overlap each other or are partially dislocated Various degrees and forms of the anomaly may occur In multiple cartilaginous exostoses and in Ollier's disease (dyschondroplasia) involvement of the lower end of the radius and ulna may produce changes very similar to those in Madelung's deformity Differ-

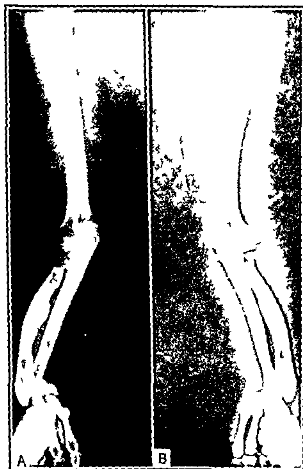


Fig 80 Madelung's Deformity Bilateral *A* Right arm *B* Left arm The changes are bilateral and similar in both forearms The radius is shortened its inferior end is deficient and tilted and there is bowing There is disturbance of the relations of the bones of the wrist

entiation in these conditions and also in the acquired types which develop as a consequence of disease or fractures of the lower ends of the bones of the forearm can be established without difficulty in most instances on the basis of the roentgen manifestations

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Developmental Considerations The development of the carapl bones follows a definite plan At birth, the human carpus is entirely cartilaginous Usually each of the carpal bones arises from a single center of ossification Common exceptions are the navicular and the capitate, either of which may arise from two or three centers It is believed that the development of an individual center of ossification for the *naviculare radiale* and a separate center for the *naviculare ulnare* results in the so-called bipartite navicular The presence of an additional center of ossification in the central portion of the bone gives rise to the much rarer condition of tripartite carpal navicular Subdivision of the navicular rather than failure of fusion of the embryonic *os centrale* with the navicular is the most logical explanation of the anomaly termed tripartite navicular The navicular is the only carpal bone which frequently arises from two or more elements The *os centrale* is a minute cartilaginous nodule There is no agreement as to the fate of the embryonic *os centrale* It has been variously reported to disappear by a process of retrograde development to degenerate and be represented in the adult carpus as a ligament that passes between the *os magnum* and the navicular and to merge with the growing cartilages of the neighboring bones into the mass of the adult capitate or the navicular

Roentgen Manifestations An adequate knowledge of the anatomical and developmental anomalies of the carpus is important in order to establish an accurate differential diagnosis between these anomalies and recent or ununited fracture The division plane of a bipartite carpal navicular is usually transverse or slightly oblique and is situated in the middle portion of the waist of the bone The roentgenogram reveals a clear space between the portions of a divided navicular and the edges are smooth and regular and usually present no arthritic changes Arthritic changes occur in the wrist joints of certain individuals with bipartate navicular who in the past have suffered undue stresses or strains in other words traumatisms which have not caused a fracture Accurate diagnosis can be established only by careful interpretation of the roentgenograms A negative history as regards trauma and similar roentgen changes in the other wrist may be the decisive factors in establishing the correct diagnosis The bone texture of each segment of a partite navicular is normal and of equal uniform density The opposing edges are smooth and sharply defined There is no evidence of osteoporosis or sclerotic changes In the presence of changes in the texture of the bone irregularity of outline or alterations in the densities at the margins of the bones the condition should be considered a fracture even though the patient can recall no injury

Partite Lunate, Bipartite Lunate

Bipartite lunate is a rare developmental anomaly The bone may be divided into two equal parts or into segments which are unequal in size The condition has been observed in two members of the same family It may be bilateral or unilateral It is not of clinical significance except that the partite lunate may be misinterpreted as a fracture The anomaly is dependent on the fact that the lunate has a double nucleus a separate bone developing from each of the nuclei The two separate segments are termed the epilunate at the ulnar aspect of the lunate and the hypolunate, which is at the radial aspect In rare instances an apparently normal and fully developed lunate may occur in association with an accessory epilunate or hypolunate

Multiple Carpal Bones

A case has been reported of a white female with fifteen carpal bones in each wrist Many of the accessory bones could be identified and named as definite previously described accessory carpal bones, others represented bipartite carpals such as the navicular and lunate Laminography was of aid to show the position location size and shape of the extra bones as well as their relation to neighboring bones of the wrist, hand and forearm

CONGENITAL FUSION OF THE CARPAL BONES

The most common of the fusion anomalies of the carpus involves the lunate and the triquetrum Less common are fusions of some of the others or of all the carpal bones There may be an associated fusion of the contiguous carpometacarpal joints These conditions are asymptomatic and may be present for years without the patient being aware of their existence The anomaly is discovered on roentgen examination and is of significance in that it may be confused with other, more important lesions Negroes are regarded as particularly liable to this anomaly The Bantus, a group of Negro people who form a native population of South Africa having migrated from equatorial Africa, exhibit this type of carpal fusion with a relatively high degree of frequency It is never a cause of symptoms There is no associated abnormality of the feet The condition is divided by Minaar into 4 subtypes (1) incomplete fusion resembling a pseudarthrosis, (2) fusion with a notch of varying depth at the site of the usual division between the two bones, (3) complete fusion of the lunate and triquetrum alone and (4) fusion of the lunate and triquetrum in association with other anomalies

Bilateral Congenital Fusion of the Carpal Capitate and Hamate

Bilateral congenital fusion of the carpal capitate and hamate is rare In the primitive digitate vertebrate there is a distal row of five carpal bones This is reduced to four as the mammal level is reached In the rabbit the fourth and fifth primary elements fuse to form one carpus which articulates with the base of the fourth and fifth metacarpals A similar state exists in certain reptiles and amphibians The human carpal bones are in an extremely primitive condition from a phylogenetic standpoint It is a matter of speculation as to whether fusion of the carpal capitate and hamate represents a phylogenetic advance It is believed that seven comprises the ancestral number of primitive digits and pentadactylism represents a reduction rather than a subdivision of one primary digit Any reduction in the number of carpal bones according to this theory constitutes a phylogenetic refinement

White reports a case in a boy nine years old with a painful tumor of the right tibia which proved to be Ewing's tumor Roentgen study of the wrists showed the centers of ossification for the capitate and the hamate approximately normal in size for the patient's age level despite the fact that these two bones were fused bilaterally The ossification of the carpus showed normal maturity except for an accessory center in the right navicular The condition is an extremely rare congenital anomaly It is asymptomatic and is discovered only during roentgen examination

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by Stockard showed that by lowering the temperature or reducing the oxygen supply at a critical time in the development of the embryo it became possible to alter the normal rate of development with the resultant production of a series of monsters and twins In his opinion, the type of abnormality is dependent primarily on the stage at which the interruption in normal development occurs rather than the specific disturbing agents Bagg's experiments with pregnant mice confirmed the hypothesis that the time rather than the agent is the significant factor

Experimental irradiation of the uterus and ovaries is known to produce abnormal offspring Mice exposed to roentgen rays prior to pregnancy and at various stages of fetal life present these changes The embryo in the uterus shows profound arrest in development, the eye and the brain particularly being affected There are also frequently defects in the limbs, the most common being club foot with dorsal flexion, syndactylism, hypodactylism congenital amputation, and polydactylism Other lesions which may occur commonly include blindness defects in the eyes and abnormalities or absence of one or both kidneys These anomalies have been observed for as long as 19 generations and appear to be inherited as mendelian recessives Other observers have thought that the disturbing factor is of less importance than the time at which the disturbance occurs Very early irradiation produces defects in the eyes Subsequently, there are defects in the brain and the branchial system Irradiation still later affects the viscera This explains the co existence of multiple anomalies affecting the eye the limb and the kidney In some instances it appears that the germ plasm undergoes alteration and the anomalies are inherited by subsequent generations It is known that irradiation of pregnant women may produce various anomalies particularly of the extremities There have been numerous experimental demonstrations that anomalies can be produced by injury to the germinal cells and that there may be transmission of the anomalies through succeeding generations Pregnant animals have been subjected to copper or riboflavin deficiency, to massive dosage of vitamin A and to selenium and lithium poisoning with the resultant production of offspring with deformed extremities Durarswami injected insulin into the yolk of chick embryos and caused anomalies which became manifested in the chickens

Many of the anomalies of the hand are inherited most probably as mendelian dominants The trait may skip a generation, alter its form of expression or otherwise behave in an atypical manner Study of human material indicates that 5 per cent of patients showing deformities of the extremities have a family history of the abnormality This small percentage may be due to several reasons It is possible that parents deliberately conceal a family history because of the feeling that it may reflect on themselves or they may be ignorant of the occurrence of the character in other members of the family In some cases this may be the first appearance of a trait that later proves to be a genetic mutation Many authorities advise against the marriage of persons with certain congenital malformations In some instances it is considered worthwhile to induce abortion or bring about sterilization However in view of the relative rarity of the anomalies it requires many generations to eliminate defects of this type and caution is advisable in this regard

There is no adequate clinical classification of anomalies of the hand and it is impossible to group many of the anomalies under definite headings Some of the congenital anomalies of the hand are not simple defects, rather

THE SESAMOID BONES OF THE HANDS

The sesamoid bones occur particularly in the regions of the joints and tendons which move the fingers and toes They are round small and flattened The two sesamoids which are usually present beneath the heads of the metacarpals are first seen in the embryo at about the third to fifth month The adult human hand as a rule has five sesamoids Two sesamoids occur at the metacarpophalangeal joint of the thumb, lying in the tendinous insertion of the flexor pollicis brevis and the adductor pollicis They blend with the ligament of the palmar aspect of the joint and articulate with the palmar surface of the metacarpal A single sesamoid is usually present at the interphalangeal joint of the thumb Sesamoids are found at the interphalangeal joints of the index and little fingers In some instances all of these are not present, while in others supernumerary sesamoids occur at the other metacarpophalangeal and interphalangeal joints

The functions of the sesamoids probably are to modify pressure diminish friction and alter the direction of a muscle pull That they are not developed to meet certain physical requirements in the adult is evidenced by the fact that they are present as cartilaginous nodules in the fetus and in greater numbers than in the adult They must be regarded as integral parts of the skeleton which are phylogenetically inherited Physical necessities probably come into play in selecting and regulating the degree of development of the original cartilaginous nodules Nevertheless irregular nodules of bone may appear as the result of intermittent pressure in certain regions *e g* the 'rider s bone which is occasionally developed in the adductor muscles of the thigh Sesamoid bones are invested by the fibrous tissue of the tendons except on the surfaces in contact with the parts over which they glide where they present smooth articular facets

CONGENITAL ANOMALIES OF THE HAND

A comprehensive report of anomalies of the hand was made by Barsky in 1951 who studied a series of 62 cases and reviewed the literature The etiology of these anomalies has not been definitely established The theory most generally accepted is that they are due to maldevelopments or mutations which are subsequently inherited A mutation is defined as a sport or permanent transmissible change in the character of an offspring as compared to the parents It was at first believed that the anomalies were due to neurogenic disturbances or amniotic adhesions during intra uterine life but this is not tenable Genetic factors are now considered the principal cause It is possible to explain the changes on the basis of conditions residing in the genes that is in the germ plasm However the cause may be transient and due to poor nutrition toxic influences and similar factors The latter group affects the fetus but not the germ plasm of the offspring In consequence the disturbance appears in one generation but not in subsequent generations It is known that pregnant mothers who suffer from measles during the early weeks of pregnancy show a high incidence of children with congenital defects such as cataracts heart disease deaf mutism and mental deficiency It is also recognized that nutritional deficiencies can affect the offspring Experiments carried on

bing in humans is most frequent between the middle and ring fingers and between the second and third toes The condition is about twice as frequent in males as in females It is often associated with polydactylism and in some families a single gene may be the cause of both syndactylism and polydactylism The condition may be associated with hypodactylism and deficiencies in the long bones Studies of the offspring of rats reared on deficient diets show that many abnormalities such as shortening or absence of the tibia fibula, radius, ulna and the bones of the hands and feet occur with syndactylism There may also be shortening of the mandible fusion of the ribs and deformities of the sternum maxilla clavicle and scapula Syndactylism may be unilateral or bilateral Short fingers occurring in addition to the syndactylism are not due to amputations Simple webbing of the third and fourth digits is usually the mildest form It may involve all of the fingers with the production of a complete mitten effect Involvement of the thumb is rare The extent of the web formation varies widely In some cases there is a shallow web while in others it extends the entire length of the joined digits In more serious cases the fusion may involve the bones the tendons and nerves A double nail at the end of the finger may also occur

Symphangilism

Symphangilism comprises a congenital fusion or absence of the interphalangeal joints, usually of the middle phalanx and the proximal or distal phalanx and occasionally accompanies syndactylism It does not occur in the simple types of syndactylism usually being associated with the more complicated forms particularly those in which extra phalanges or brachydactylism are present The condition is inherited or inheritable The lesion does not lend itself to surgical cure in most instances Cushing in 1916 studied a large family with this condition He stated that the middle phalanges are the last group or row of phalanges to ossify and this accounts for their relative shortness and tendency to fuse Where less than four fingers are affected the fusion occurs in those farthest from the index finger It appears that the trait may be transmitted from a parent in whom it is inconspicuous but never by parents who are not affected In the family studied by Cushing the condition had been transmitted through seven generations and of 302 individuals comprising 72 families for whom full records were available 25 8 per cent had been affected Of 150 children of affected parents 78 or 52 per cent showed the anomaly The trait thus behaves as a simple mendelian dominant with an equal chance that it will or will not be inherited The distal joints of the fingers and the toes may be affected The fusions may involve the talus the navicular and other bones of the wrist and tarsus

Polydactylism

Polydactylism is a part of the general phenomenon of duplication of parts of the body The condition may vary from complete duplication of a limb to doubling of a single phalanx It occurs in many animals and fowl as well as humans tends to be inherited and frequently accompanies syndactylism brachydactylism brachyphalangism or other congenital anomalies The lesion is usually marginal that is the accessory

are complicated and combine a number of related or unrelated malformations It is difficult to decide the main category in which certain cases should be classified It is necessary in discussing these anomalies to consider each one individually Muller has published a monograph in which he discusses the congenital hereditary and constitutional deformities which have been observed in the human hand These are so varied and numerous that it is impractical to classify and codify them Each example forms a stone in a mosaic which can be understood only when it is viewed in relation to the whole At present many blocks necessary to complete the pattern are missing Each new case is important in that it helps complete the design If a malformation of the embryonic body occurs before the skeleton has developed the skeleton adapts itself to the abnormal configuration of the part of the body in which it is placed Lack of a part of an extremity or of the entire extremity causes aplasia of some or all of the bones of the extremity In cases of polydactylism the supernumerary digit of the hand or foot develops in the bud stage of the extremity Defects of the neural arches of the vertebra termed spina bifida are the consequence of primary disturbances in the development of the spinal cord

The wide variety of anomalies of the hand that may appear at birth are best grouped into those that are inherited those caused by noxious influences during pregnancy and those due to atavism or regression to a more primitive form In 1905 Farabee demonstrated that in a family with brachydactylism certain anomalies are controlled by inheritance The most striking example of a mendelian dominant pattern is Hegdekatti s report of six generations of an Indian family in which the malformation comprised a reduction of the digits to one in all four extremities In 1917 Drinkwater recorded a family with symphalangism dating from the time of Henry VI of England According to legend a member of the family had first been noted to have the anomaly in 1453 In 1874 when a tomb was opened during the course of construction on church grounds the finger bones showed anomalies similar to those found in direct descendants living in a nearby town Examples of congenital malformations originating as mutations and subsequently inherited along mendelian lines have been recorded in the literature with increasing frequency

In the case of hyperphalangism atavism or inheritance of characters from remote ancestors in the phylogenetic line may be important in etiology To account for hyperphalangism on such a basis however one must revert to the phalangeal formula of the reptile since no animal higher than the reptile has more than three phalanges in a single finger or toe nor more than two phalanges in the thumb or great toe Leboucq assumed that the anomaly was due to a process of budding with an extra phalanx developing from the proximal phalanx Pol was of the opinion that the supernumerary phalanx developed as an independent growth of the proximal epiphysis of the proximal phalanx whereas Grafenberg stated that the extra phalanx developed from an additional proximal epiphysis of the terminal phalanx

Syndactylism

Syndactylism fusion or absence of the digits is the most common congenital anomaly of the hand and occurs about once in every 3000 births It has been described in gibbons marsupials and other mammals Web-

does not function The condition may occur as a solitary anomaly or in conjunction with annular grooves and syndactylism While the digits are usually separate, there may be slight webbing of the proximal phalanx In the case of brachydactylism associated with syndactylism, the web can be eliminated With a flail joint, arthrodesis results in a stable fingertip In the presence of symphangilisms, no treatment is required

Combined Anomalies

McGavack and Reinstein report a case of a forty year old married female who was born with hands and feet which were small and unusual in appearance She left school at the age of sixteen years, while still in the fourth grade of grammar school During the previous four years, she had gained weight from 127 to 187 pounds and developed hairiness of the chin and high blood pressure Her two children, eight and twelve years old respectively, presented no physical deformity She had a very obese sister and a brother s daughter, both of whom showed deformity similar to that of the patient The patient had a hypopituitary habitus and a slight degree of microcephaly Both hands were small and showed symmetrical variations from the normal All of the fingers were short This was most noticeable bilaterally in the first second, and third, fingers Neither the index nor the middle finger was as long as the little finger and both presented abnormal ulnar deviation The distal phalanx of the fifth finger had an inclination to the radial side of the hand Roentgen study revealed (1) abnormal shortening of the first, fourth, and fifth metacarpal bones, (2) enlargement of the base of the first phalanx of both index fingers with radial displacement On the right side this digit had the appearance of a metacarpal bone, (3) shortening and broadening of the bases of the phalanges of the middle fingers and the left index finger, (4) polyphalangism of both middle and the left index fingers with the interpolation of a short, broad bone between the proximal and middle phalanges in each instance (5) marked shortening of the middle phalanges of all the fingers, (6) ulnar deviation of the bones of the index and middle fingers, and radial deviation of the last phalanx of the little fingers The feet were small as compared with the legs The toes were very short, otherwise showed no changes This was confirmed by roentgenograms in which the middle phalanges of the second, third and fourth toes were found to be exceedingly short and broad The middle phalanges of the little toes were missing and the base of the distal segment broadened

Three degrees of maldevelopment of the hand may be recognized (1) deformities believed to result from abnormal skeletal anlages (2) disturbances due to failure of full development of the primitive hand plate in which the preceding stages have been normal and (3) abnormalities which appear in the evolution of the hand from an apparently normal primitive stage The striking feature in the case described is the presence of the brachydactylic triad of brachymesophalangism hyperphalangism, and shortening of the first metacarpal This is a rare combination despite the fact that many instances of the condition have been unreported The common denominator of the group lies in the shortening and broadening of the proximal and middle phalanges of the second and third fingers and the interpolation of at least one additional phalanx It appears that no two patients present exactly the same or identical deformities In three children of the group supernumerary epiphyses have been present in two

digit is present on either the ulna or the radial side of the hand rather than centrally The common form is hexadactylism Duplication of the thumb is more common than duplication of the little finger Radial and ulnar polydactylism may occur together in the same family although they are probably determined by separate genes The anomaly occurs in three main types In the first, there is an extra mass which is not adherent to the skeleton and frequently contains no bones cartilage muscles or tendons The second type consists of duplication of the digits with formations similar to an ordinary finger or toe which contain bones and articulate with an enlarged or bifurcated metacarpal The third form is rare and comprises an extra digit with its own metacarpal or metatarsal bone The anomaly may be unilateral or bilateral While the common form consists of one extra digit, there are many instances with seven toes or seven fingers and a case has been reported with 13 fingers on each hand and 12 toes on each foot The bifid thumb comprises a form of polydactylism in which the duplication is limited to the terminal phalanx The duplication of the distal phalanx occurs most commonly in the thumb but may also affect other fingers In some instances the extra phalanx has been considered an exostosis The lesion is important in that it may seriously impair the function of the thumb Correction by surgery in adult life is difficult and in most instances proves unsatisfactory In children however correction of the deformity is possible without jeopardy to function, and when the diagnosis is established in infancy or childhood the accessory ossicle should be removed As the ossicle does not normally serve for the attachment of muscles or ligaments its ablation does not alter the function of the hand The sooner it is removed the less the danger of laxity of the capsule or damage to the epiphysis The operation is simple and the defect is equalized by normal epiphyseal growth The roentgen picture varies widely The head of the first metacarpal may be irregular and wedge shaped and articulate with bifid proximal phalanges which are united at their proximal extremities The bifid phalanges may diverge at a right angle or a lesser or greater degree of angularity Another form of the malady consists of interposition of a small wedge shaped bony mass between the proximal and distal phalanges of the thumb The supernumerary wedge shaped bone between the proximal phalanx and the epiphysis of the distal phalanx may cause radial deviation of the distal phalanx

Brachydactylism

Brachydactylism indicates shortening of the fingers The embryonic development of the fingers is affected in such a manner that one several or all of the digits become shortened This may be due to a decrease in the length of the phalanges a diminution in the number of phalanges or shortening of the metacarpal It frequently occurs in conjunction with webbing of the fingers polyphalangism or polydactylism Brachydactylism is the prime example of mendelian inheritance demonstrated in man and is believed to be a simple dominant In some cases the fingers function normally the profundus tendon being attached to the distal phalanx and being able to flex the finger In other cases there is no attachment of the muscle to the terminal phalanges with the result that the distal interphalangeal articulation is flail like In consequence the patient is required to assist the distal phalanx with the other fingers in order to make a pinching motion In some cases the joint is rigid and

is that the condition is due to genetic factors Many cases of intra uterine amputation demontsrate that sharply circumscribed areas of limb bud tissue are of such inferior quality that imperfect histogenesis has occurred The condition is often referred to in the literature as Streeter's dysplasia

There are many examples of amputation of the digits and larger portions of the extremity due to germinal effects At the time of birth, only a depression, groove, or healed stump with residual strands may remain visible The lesion may be bilateral or unilateral It is important to conserve rudimentary digits The lesion may consist of absence of the hand, either unilateral or bilateral With complete absence of the hand, there may be merely a soft tissue mass at the distal end of the forearm The lower third of the legs and the feet may be absent unilaterally or bilaterally In some instances the patient learns to write and carry on other functions with the stump

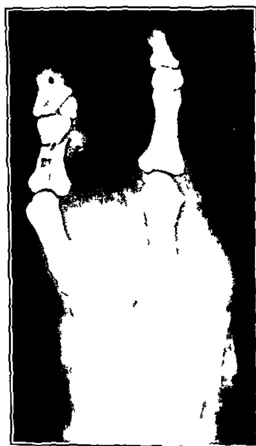


FIG 82 Congenital Anomaly of Hand Cleft Hand The thumb and one additional digit are present The phalanges of both these fingers and the carpus show multiple anomalies

Cleft Hand or Lobster-claw Hand

In this rare congenital anomaly there is a defect involving the central portion of the hand, the digits being divided into a radial and an ulnar group The changes may be bilateral or unilateral The condition appears to be definitely hereditary There are several forms of cleft hand The first type is characterized by an extensive V shaped cleft which tapers and divides the hand into two segments The digits in each portion are often webbed The middle metacarpal and finger are usually missing in which case the hand resembles a claw In another variety of the same

cases and there was absence of normally appearing epiphyses in two. Incomplete longitudinal bony division or notching of the proximal and middle phalanges has been described. It appears that the case described above presents a higher degree of disturbance of the formation of the anlage than others in the same general group. Supernumerary phalanges occur in the second and third digits only. In Leboucq's report it was recorded in both hands. In one of the cases reported by Joachimsthal, they are present in both fingers of the right hand and the left index finger.

Annular Grooves and Congenital Amputations

The lesion may comprise a very shallow groove in the skin or the subcutaneous tissue affecting only one digit or there may be multiple deep grooves extending almost to the bone with a small distal segment of the finger at-



Fig 81 Congenital Amputations and Annular Grooves. There are congenital amputations of the fingers with annular grooves at the proximal ends of the index, middle and ring fingers. The patient is six months of age and presents no other congenital anomalies.

tached by a pedicle. Annular grooves may also occur in the foot, leg and forearm, often in conjunction with syndactylism and brachydactylism, the defect usually being combined. There may be a groove on one hand while the other hand may show syndactylism and partial absence of the distal portion of a phalanx. Shallow rings do not interfere with the function of the finger. Deep or multiple rings frequently result in edema, in which case the groove must be excised to the level of the normal structures and the subcutaneous tissues *approximated in order to prevent an annular constricting scar*. The etiology of uterine amputations is not clear. They have been considered due to gangrene, inflammatory changes, diet, toxic factors such as measles and the Rh factor. In the earlier literature these were believed to be caused by amniotic bands. However, the belief now

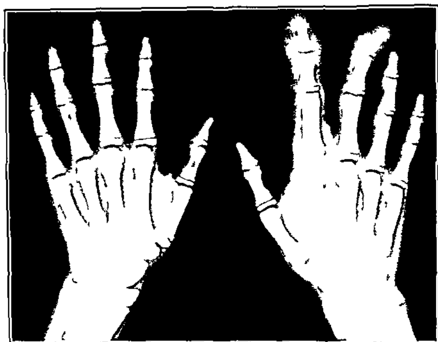


FIG 83 Megalodactylism There is a developmental anomaly with marked elongation and increase in the width of the index and middle fingers of the left hand The soft tissues of the affected fingers are widened and denser than normal

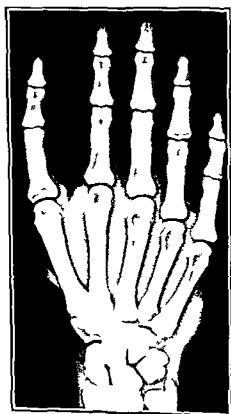


FIG 84 Five Fingers with Absence of Thumb

general type, the cleft is shallow and the second metacarpal is present but the digit is missing These anomalies are usually bilateral and often are associated with similar deformities of the feet Another type of cleft hand is characterized by the presence of a solitary radial digit or thumb with its metacarpal and an ulnar digit, the remaining digits and metacarpals being absent This type of cleft hand is generally unilateral and there is no associated deformity of the foot The treatment of lobster-claw hand varies with the character of the deformity In the presence of a deep cleft, elimination of the cleft is usually advisable However, if function is good and there is no great advantage in merely improving the appearance, operation is inadvisable In cases with only an ulnar and radial digit the grasp of the hand may be improved by releasing the web with a Z shaped incision

Megalodactylism

Hypertrophy of the digits is termed megalodactylism or macrodactylism The second and third digits are most apt to be involved and the condition is more common in males than in females It is rarely hereditary and the etiology has not been explained In true hypertrophy there is enlargement of all the elements of the finger This may occur in the presence of an angioma The only treatment is partial amputation to overcome the unsightliness of the condition When due to a neurofibroma or an angioma removal of the excessive tissue results in improvement of the condition

Miscellaneous Anomalies

There may be congenital absence of the long extensor muscles of the thumb and other tendons and muscles There have been cases described with five fingers in each hand but no thumb (Fig 84) Cases may occur with seven or six toes and in some the condition has been hereditary being present in both mother and child

The Laurence-Moon-Biedl Syndrome

The Laurence Moon Biedl syndrome is characterized by obesity, atypical retinitis pigmentosa mental retardation polydactylism genital hypoplasia and familial occurrence There are six fingers on each hand The supernumerary finger may be composed of one or more phalanges with a fifth metacarpal bone which is bifid abnormal in size or otherwise malformed Anomalies of the toes may also occur with variations in the number size and shape of the phalanges

Hyperphalangism (Polyphalangism) and Brachydactylism Hereditary Abnormal Segmentation of the Hand

Hereditary abnormal segmentation is a syndrome that involves the hands and feet and is characterized by more than the normal number of phalanges in the affected finger This feature of the syndrome is called hyperphalangy or polyphalangy and should be distinguished from polydactyly in which there are more than the usual five fingers in the individual hand No definitive or etiologic name has been applied to the syndrome

a third the normal length), the proximal phalanx of the ring finger is abnormally long, the base of the proximal phalanx of the index finger articulates with the metacarpal head obliquely and deviates ulnarward, and in preadolescent patients there is an extra, roughly triangular bone at the base of the index finger that probably corresponds to the extra phalanx in the middle finger (this supernumerary bone usually fuses with the proximal phalanx during adolescence) The hand is grossly deformed and the anomaly is obvious on casual examination The index, middle and little fingers are short and stubby, being equal in length to the thumb whereas the ring finger is unusually long There is an extra flexion crease in the middle finger The interphalangeal joints show hypermobility, with a range considerably greater than normal

Although rare, this syndrome is important It comprises a series of anatomic abnormalities inherited along mendelian dominant lines A patient with these anomalies should be told of the likelihood of their appearance in his offspring Before marriage, the possibilities of transmission of the syndrome should be explained, since failure to do so may result in medico-legal complications Many states require premarital examination of both participants, and it is unethical to withhold information about contagious or inheritable disease It is important to recognize the disease when such a patient s hands have suffered an injury The supernumerary bones in the index or middle fingers may be confused with old ununited fractures or synostoses The clinician should be aware that there is a tendency to an increased number of congenital anomalies in other parts of the body, and survey roentgenograms of the skeleton are essential to determine their location and significance There is no interference with function because of the deformities, and the patients do not appear to be hampered in manual pursuits

Dystrophy of the Fifth Finger

Wilson reports a rare deformity of the fifth finger characterized by clubbing similar to that in the fingers of persons with chronic pulmonary disease and a typical radial deviation of the terminal phalanx The abnormality was first described by Kirner in 1927 who ascribed the condition to osteochondritis Brailsford reported a case in a girl age nine He stated that the radiographic changes were characteristic and comprised increased density of the metaphysis while the distal portion of the epiphysis was ragged and concave In 1944 Mercer described a case in a girl with bilateral changes Wilson found four cases during a period of three years The clinical appearance in each was identical and characteristic Roentgen studies showed that the deformity of the terminal phalanx of the little finger was due to deviation of the metaphysis with relation to the epiphysis the deviation being anteriorly and to the radial side The cause of the condition is not known It is believed to be a congenital anomaly The age of onset is usually between two and eight years There is a history of gradual development of the deformity over a period of six to twelve months, which does not support the theory of a congenital origin There is a familial tendency in some instances The age incidence and the roentgen appearance suggest that the lesion is an osteochondritis The deformity of the terminal phalanx may be caused by two factors The long flexor is a more powerful muscle than the extensor and can produce anterior bending during the active phase of

Historical Survey Leboucq recorded the first case of this syndrome in 1896 Vidal, in 1910, described a family tree of five generations with ten cases physically and roentgenographically similar to Leboucq s case The condition acted as a mendelian dominant, although in certain persons the expected deformity did not occur Drinkwater's report (1916) of four generations of a family is the first account of this syndrome in the English language He found eight males and eight females affected in 43 members of the family Dell'Oro, in 1935, recorded one case The patient was a boy who had been brought to the emergency ward after an automobile accident X ray study of the extremities was made and after preliminary examination he was thought to have a fracture of the proximal phalanx

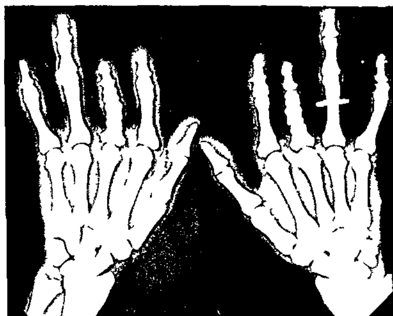


Fig 85 Hyperphalangism The middle finger of each hand shows a supernumerary phalanx the combined length of the 2 proximal phalanges being less than that of the proximal phalanges of the other fingers The middle phalanges of the index and ring fingers are small and abnormal in shape *The first metacarpal bones are decreased in size The proximal phalanx of the ring* finger of each hand is elongated The proximal phalanges of the index finger are long and present a notch like defect at the proximal ends with transverse linear bands of increased density at the sites of the previous epiphyseal lines The ring fingers are elongated The little fingers are deviated toward the center of the hand

of the middle finger This was later proved to be the extra interphalangeal articulation between the supernumerary phalanx and the true proximal phalanx In 1937 Lossen found ten cases in the German literature and added one of his own Becker in 1939 described a family with multiple congenital anomalies of the hand among which there were several cases of hyperphalangism Fifteen cases during five generations were described In 1941 McGavack presented the case of a forty year old married woman whose sister and niece showed similar deformities of the hands

Roentgen and Clinical Manifestations The syndrome consists essentially of five anatomic features hyperphalangism (polyphalangism) with four rather than three phalanges in the middle finger and sometimes the index finger brachyphalangism or shortening of certain phalanges (the middle phalanx of each finger and toe is markedly shortened being about

It does not occur in rickets, osteomalacia and osteogenesis imperfecta, in these conditions there being bending of the femurs and pelvis without involvement of the acetabulum Otto's pelvis is most commonly encountered in the fifth to seventh decades of life It may develop in the late stages of many diseases of degenerative rheumatoid and pyogenic origin and in young persons after severe trauma or chronic infections The condition does not develop in recumbent individuals and after the inflammatory changes have subsided It is much more common in females In the active phase of the disease the acetabular walls are fragile and fracture easily The new bone formation along the walls of the protruding acetabulum shows increased density During childbirth the protrusion may form a serious obstacle to normal delivery



Fig 86 Intrapelvic Protrusion of the Femoral Heads Otto s Disease There is intrapelvic protrusion of the heads of the femurs The hip joint spaces are narrowed There are extensive cystic changes involving the heads of the femurs and the right acetabulum

osteochondritis The natural position of the little finger in flexion lends itself to radial and anterior displacement, especially since the little finger points toward the radial border of the wrist and the ulnar border of its terminal phalanx lies in relation to the hypothenar eminence Lateral resistance to the flexed finger is from the ulnar aspect This tends to produce a radial deformity The roentgen manifestations of increased density in the terminal phalanx in the early stage with return to normal density in the later stages are in conformity with the changes expected in osteochondritis Osteochondritis of the finger usually affects the epiphysis and in the reported cases the roentgen changes are in this region

ANOMALIES OF THE PELVIC BONES ASSOCIATED WITH EXSTROPHY OF THE BLADDER

Exstrophy of the bladder is a congenital anomaly characterized by absence of the anterior abdominal wall and the anterior wall of the bladder There is consequent exposure of the ureteral orifices and the posterior vesical wall The roentgen diagnosis may be made by the demonstration of changes in the bones of the pelvis which occur in association with this anomaly The bones of the pubis are small maldeveloped *and widely separated in some instances being separated by the* width of the sacrum There is marked flaring of the wing of the ilium bilaterally resulting in a pelvic inlet which is square rather than round or oval as normally Similar anomalies in the bones of the pelvis may occur in epispadias The treatment is usually transplantation of the ureters into the sigmoid

INTRAPELVIC PROTRUSION OF THE ACETABULUM OTTO PELVIS

The Otto pelvis intrapelvic protrusion of the acetabulum consists of deepening of the hip joint central protrusion of the head of the femur into the acetabulum and bulging of the acetabulum into the pelvis The underlying cause appears to be a congenital or developmental defect since in many cases there is symmetrical involvement bilaterally without apparent underlying cause In some instances the onset of the condition is observed at about the time of puberty It is believed that a *form of* growth disturbance incidental to puberty and weight bearing are the significant underlying factors Trauma and local inflammatory changes appear to play a part in certain cases Although a definite familial tendency has been recognized a true hereditary tendency has never been established The condition may be classified into two main groups (1) the idiopathic type which is bilateral *more common in females* and occurs at or about puberty and (2) the acquired form which is frequently unilateral and shows considerable sclerotic change in the bones about the hip joint The walls of the acetabulum are displaced medialward envelop the femoral heads and push the lateral wall of the pelvis inward The condition was first described by Otto It is due to pressure exerted on the acetabulum while it is pliable and soft The pressure is caused by the weight of the body applied to the hip joints in the upright position in individuals whose femoral heads are more resistant than the acetabulum

and without it the lower extremity loses its optimum muscular and mechanical efficiency In the embryological development of this bone the cells which are destined to become the early patellar nucleus are imbedded in the substance of the developing quadriceps muscle mass The embryonic state of these structures makes their relationship very close The synovial cavity of the knee appears first and develops more rapidly on the medial side Similarly the development of the patellofemoral joint space is completed earlier on the medial aspect Since the patella develops in relation to the condyles of the femur and passes through a stage of fusion with the condyles the medial and lateral areas of the articular sur-

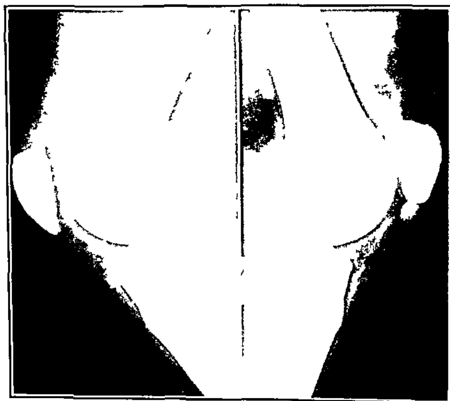


FIG 87 Partite Patella The patella is divided into two portions the lower fragment being much smaller than the upper The division between the two portions is sharply defined and smooth in outline

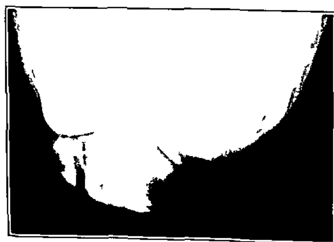


FIG 88 Partite Patella
The division of the patella is clearly visualized

The roentgenographic findings are those of deepening and markedly increased density about the margins of the acetabulums, inward protrusion of the lateral pelvic walls and envelopment of the heads of the femurs by the deepened acetabulums There is flattening mottling, eburnation and irregularity of outline of the weight-bearing portion of the head of the femur The hip joint spaces become narrowed but are not obliterated There is spurring about the margins of the acetabulums and the heads of the femurs Osteoporosis of the bones of the pelvis and the femurs is usually present

CONGENITAL COXA VARA

Congenital coxa vara is relatively rare and of unknown etiology At birth, the upper end of the femur is composed entirely of cartilage The neck becomes ossified at the fourth year with the exception of the epiphyseal line A disturbance of ossification of the neck takes place during the first four years of life prior to the ossification of the neck After the fifth year of life the lesion is definitely demonstrable by roentgen methods The femoral head is displaced downward and a vertical fissure appears lateral to the epiphysis and separates it from the remainder of the neck The segment which presents the appearance of the head is in reality the anatomical head and the epiphyseal cartilage with a triangular fragment of the neck The descent of the head due to a defect in the femoral neck comprises the fundamental pathologic lesion and in many instances is accompanied by shortening of the femur The condition is characterized by a painless limp which first becomes noticeable as the child begins to walk In later years there is pain in the hip Physical examination reveals shortness of the extremity and a waddling gait The greater trochanter is elevated and prominent The principal condition which must be considered in differential diagnosis is congenital dislocation Roentgen examination establishes the true nature of the lesion The diagnosis is not difficult if the possibility of this condition is borne in mind The femoral neck is bent and the head is depressed The limb is maintained in adduction The epiphyseal line is more vertical in position than normally and appears to be branched like an inverted Y The head of the femur is large, shows increased radiance and lies at the inferior aspect of the acetabulum The appearance closely simulates a fracture of the neck of the femur However fracture is extremely rare in children The neck is imperfectly formed and there is a zone of rarefaction which contains osseous nuclei and demarcates the triangular fragment During adolescence the abnormality is usually considered to be an ununited fracture As the treatment of these conditions is the same an error in diagnosis causes no serious consequences The therapy consists of subtrochanteric osteotomy with fixation to alter the axis of weight bearing in the neck of the femur and facilitate ossification

PARTITE PATELLA

The patella has long been a controversial subject of study among students of anatomy and embryology One school considers it a structure which is slowly eliminating itself as an essential part of the human skeletal framework and believes that the patella impairs the performance of the quadriceps extensor apparatus Others state that the patella is essential

He suggests that prenatal mechanical molding of the bones and pressure atrophy of the fetal skin is the probable pathogenic mechanism and stresses the fact that the changes in the legs are similar to those caused by prenatal bowing in rickets and Blount's tibial osteochondritis In each of the cases there was congenital, symmetrical bowing of the femoral and humeral shafts and similar but not necessarily paired deformities of the shafts of the tibia, the radius and the ulna bilaterally The bowed diaphyseal segments were thickened and bent All of the curvatures extended through substantial longitudinal segments of the shaft in or near the middle third Large symmetrical cutaneous dimples overlay the salient angles of the curves in several of the deformed long bones There was no evidence of intrinsic prenatal skeletal dysplasias such as achondroplasia or dyschondroplasia Also, there were no manifestations of fragilitas ossium (osteogenesis imperfecta) pseudarthrosis, prenatal or infantile rickets or syphilis The bowing may be mistaken for that in rickets and it is probable also that cases which have been reported as instances of infantile tibial osteochondrosis were in reality examples of prenatal bowing of the bones of the legs In many instances the deformities disappeared with advancing age, particularly in the upper extremities In other cases the osseous defects remained unchanged until the third year of life The dimples persisted during infancy without significant change with advancing age

CONGENITAL GENU RECURVATUM CONGENITAL DISLOCATION OF THE KNEE

Genu recurvatum or congenital dislocation of the knee often occurs in association with other congenital abnormalities especially clubfoot and congenital dislocation of the hip The patella may remain undeveloped until the deformity has been corrected surgically and normal functional stresses restored McFarlane reports 4 cases in a colored woman and her three children Each of the children had a different father and none of the fathers had any congenital abnormality All the grandparents were normal as far as could be ascertained The first patient was a girl aged ten months with bilateral congenital genu recurvatum Reduction was accomplished by open operation When the child was last seen, eight years later both knee joints showed normal function and range of motion The second patient was a boy of seventeen months Both knees were operated on successfully and after two years he had a normal range of movement The third child a girl, was seen at the age of three years with bilateral knee recurvation and severe gastroenteritis X ray examination showed a typical deformity of the knees The infant died before treatment could be given The mother, twenty seven years old had untreated bilateral genu recurvatum Roentgen study showed complete posterior dislocation of the knee

CONGENITAL BOWING OF THE TIBIA CONGENITAL KYPHOSCOLIOTIC TIBIA

Congenital kyphoscoliotic tibia is also known as congenital bowing of the tibia and intrauterine fracture of the tibia While the deformity comprises principally an anterior or posterior bowing of the tibia associated

faces are at first approximately equal in size A change in the relative size begins after the patella separates from the femur This occurs slowly and at the 192 mm stage the articular surface of the patella is divided by a vertical ridge into a large lateral and smaller medial area which are relatively comparable in size to those in the adult

The common developmental aberration of the patella due to pathological genes is manifested by faulty and incomplete differentiation of the pre-muscle mesenchymal mass which is destined to become a portion of the quadriceps extensor apparatus, the vastus internus The medial portion of the knee joint possesses older phylogenetic structures than its lateral portion Anomalies in the medial compartment will present themselves in the form of developmental erasure Anomalies in the lateral portion will present themselves in the form of accessory growth Accessory patella centers are constantly found in the lateral portion The most common type of bipartite patella is characterized by the smaller fragment lying in the upper and outer quadrant of the bone Other types are those with transverse and vertical fissures the emarginate variety in which there is actually no second fragment, and the multipartite type The characteristic manifestation on the roentgenogram is the presence of a smooth, sharply defined space between the fragments There is no irregularity of outline or swelling as in fracture

CONGENITAL ANOMALIES DUE TO FAULTY INTRAUTERINE POSITION

Numerous malformations may occur as the result of faulty position during intrauterine life These are characteristic and can be identified with definiteness Infantile bowing of the legs and skin dimpling may be the result of intrauterine pressure of the opposite foot upon the mid portion of the thigh Skull indentations may result from the pressure of a hand or foot during the later months of intrauterine life Faulty postural habits established before birth may persist A common example is persistent adduction of one femur with contraction of the adductor muscles elevation of the ipsilateral pelvis and a contralateral scoliosis in the lumbar region The abnormality of growth may continue and the abnormal stresses exerted through the adducted femur to the acetabulum may result in underdevelopment of the acetabulum and obliquity of its roof Correction of the adduction contracture and application of the proper stress to the acetabulum results in prompt disappearance of the dysgenesis with return of the acetabulum to normal While primary dysplasia of the acetabulum is believed to occur the above mechanism is an adequate explanation of this abnormality and may constitute the sole cause of the condition

PRENATAL BOWING AND THICKENING OF TUBULAR BONES WITH MULTIPLE CUTANEOUS DIMPLES IN ARMS AND LEGS A CONGENITAL SYNDROME OF MECHANICAL ORIGIN

Caffey reports the clinical and roentgen findings in three cases characterized by prenatal bowing and thickening of the tubular bones in young children associated with multiple dimplings of the skin of the extremities

teristic picture is frequently not present, in some instances the defect consisting of only absence of the proximal epiphysis of the fibula while in others there are severe bilateral deformities of the legs with congenital changes in other parts of the body. The defects occur in the embryo prior to the sixth or seventh week of fetal life. The cause is unknown. Middleton suggests that absence of the fibula may be secondary, the primary abnormality being a failure of maturation characterized by diminution of growth in the length of the muscle fibers; failure of longitudinal growth of the muscle resulting in a relative shortening of the calf and peroneal muscle and placing an abnormal stress on the tibia and foot with resultant

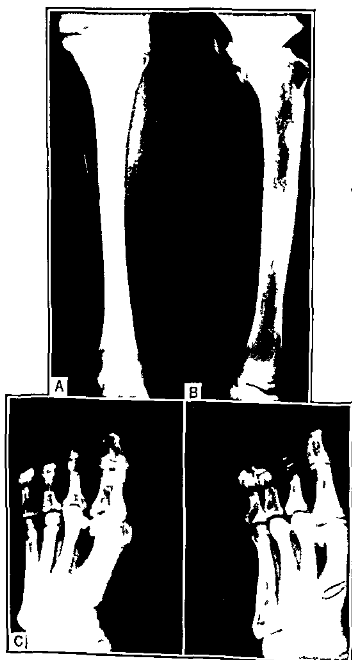


Fig 89 Congenital Absence of the Fibula Associated with Four Rays in the Foot. *A* Antero posterior view lower leg. *B* Lateral view. The fibula is completely absent. *C* Antero posterior and oblique views of the foot showing only four rays, the fifth metatarsal and the little toe being absent

with a lateral or medial rotational deformity, in rare instances the deformity is entirely or chiefly in a single plane Because of the similarity to deformities of the spine, the term kyphoscoliosis is particularly applicable The lesion is characterized by certain specific features which differentiate it from the bowings due to other causes the changes being confined to the distal portion of the middle third of the bone with an ossified mass at the site of the deformity There is a potentiality of spontaneous correction by normal epiphyseal growth The condition frequently occurs in association with a characteristic group of deformities which include hypoplasia of the fibula and the bones of the foot, syndactylism gigantism and skin lesions There is dimpling of the skin at or near the apex of the curved tibia

The etiology is unknown The condition may be due to a congenital defect of ossification, a hereditary fault or abnormal intrauterine pressure There is usually retardation of growth of the affected tibia which results in a significant inequality of length The condition is similar to that described by Caffey and termed prenatal bowing and thickening of tubular bones, and in some cases the humerus and femurs are involved as well as the tibia It is not related to congenital pseudarthrosis as no defect in the bony structure has ever been demonstrated

Congenital bowing of the tibia may occur in various forms (1) with anterior bowing (2) with anterolateral or anteromedial bowing (3) with posterolateral or posteromedial bowing (4) kyphoscoliotic tibia with cystic areas the prepseudoarthritic type and (5) kyphoscoliotic tibia with pseudarthrosis In cases characterized by bowing with anterior or posterior convexity the diaphyseal segment is principally affected Delay in ossification of the lower tibial epiphysis is *frequent although normal* development eventually occurs Spontaneous correction is particularly apt to ensue in instances of posterior bowing Pathological fracture is common and is associated with pseudarthrosis due to non-union In cases with anterior bowing the thickening is most marked at the site of maximum curvature and extends practically to the anterior cortex of the tibia The bony density decreases gradually and progressively toward the epiphyseal ends of the diaphysis In cases with posterior and medial bowing there is an associated talipes calcaneovalgus and the tarsus appears to nestle in the concavity of the tibia This type has been described in the literature as intrauterine fracture of the tibia and fibula and constitutes an important clinical entity The calcaneovalgus deformity can be corrected without difficulty The posterior angulation responds well to gentle molding and fixation in a cast The medial bowing is more resistant to treatment

CONGENITAL ABSENCE OF THE FIBULA

Of all the long bones the fibula is the one which is most commonly congenitally absent Congenital absence of the tibia the ulna the radius and the femur occurs less frequently The diagnosis is established by roentgen examination The classical syndrome comprises partial or total absence of the fibula anterior bowing of the distal third of the tibia with a dimple in the skin at the point of most marked angulation marked talipes equinovalgus absence of one or more rays of the foot and lack of one or more or fusions of two or more of the tarsal bones The charac

SESAMOID BONES OF THE LOWER EXTREMITY

Sesamoid bones are found occasionally in the tendon of the gluteus maximus as it passes over the greater trochanters, in the tendon of the psoas major, where it glides over the pubis, and in the lateral head of the gastrocnemius, behind the external condyle of the femur In the lower extremity the largest sesamoid bone of the joints is the patella, developed in the tendon of the quadriceps femoris Sesamoids are also present in several of the tendons of the lower limb, *viz*, the tendons which wind around the medial or lateral malleoli, the tendon of the peroneus longus, where it glides on the cuboid, the tendon of the tibialis anterior, opposite the smooth facet of the first cuneiform bone, and the tendon of the tibialis posterior, opposite the medial side of the head of the talus

The sesamoids of the feet are found in the lumbrical and rarely the interosseous tendons in the region adjacent to the joint surfaces Two sesamoids are present in practically all individuals at the metatarsophalangeal joint of the great toe Each of these sesamoid bones is enclosed in the tendon of the flexor hallucis brevis and the articular capsule They lie in close relation to the head of the first metatarsal and articulate with



Fig 90



Fig 91

Fig 90 Congenital Absence of the Fibula On the left the fibula is absent the tibia is shortened bowed and dense and the femur is decreased in size

Fig 91 Accessory Tarsal Navicular
The accessory tarsal navicular is sharply outlined and smooth in outline (arrow)

bowing and equinovalgus Heredity plays a minor role and no familial instances of absence of the fibula have been reported In most cases the pregnancy and birth have been normal None of the patients' mothers have had German measles during pregnancy In a case reported by Coventry the lesion occurred in one of twin boys the other twin being entirely normal The condition is more commonly unilateral than bilateral There appears to be a slight predilection for males

Classification The condition has been divided into three principal groups Type 1 includes partial unilateral absence of the fibula with little or no bowing of the tibia The degree of absence may vary from involvement of the proximal epiphysis to almost complete absence of the bone The extremity is usually shortened Deformity of the foot is slight or absent Congenital anomalies do not occur in other parts of the body This form frequently gives no clinical manifestations and is usually discovered unexpectedly during roentgen examination In type 2 the fibula is entirely or practically completely absent The lesion involves only one side There is deformity of the leg with anterior bowing of the tibia dimpling of the skin, equinovalgus of the foot and absence or deformity of the rays and tarsal bones The extremity is markedly shortened Type 3 is characterized by absence of the fibula associated with other congenital anomalies or bilateral absence of the fibula The associated anomalies may involve the upper or lower extremities the femur often being anomalous The femur on the side of the defect is usually shortened as the development of the capital femoral epiphysis and the acetabulum is delayed Spina bifida may occur

Differential Diagnosis The condition may be mistaken for congenital absence of the tibia The foot and ankle in congenital absence of the tibia are held in varus not valgus and the medial malleolus is missing with resultant displacement medialward of the ankle In congenital absence of the tibia there is little or no anterior bowing of the leg but dislocation of the knee does occur because of absence of the normal articulation of the tibia with the femur It may be difficult by roentgen methods to distinguish between the tibia and fibula The deformity of the leg aids in establishing the diagnosis The fact that the upper end of the fibula is not broadened and does not form a true articulation with the femur helps to distinguish it from the tibia Congenital pseudarthrosis of the tibia may produce an appearance similar to that in congenital absence of the fibula Other congenital anomalies are rare in association with congenital pseudarthrosis but are common in congenital absence of the fibula In congenital pseudarthrosis the tibia and fibula are thickened and may be angulated or fractured Shortening of the lower extremity without absence of the fibula may occur but this condition can easily be differentiated by roentgen examination

Treatment It is recommended that treatment be instituted as soon after birth as possible The position can be corrected at least partially by the use of casts In some instances surgical correction may be delayed until the child begins to walk at which time a prosthesis is made The child is permitted to use this prosthesis until the age of two to three years, after which time operative procedures are performed Amputation is seldom necessary and should not be performed in bilateral absence of the fibula In cases of unilateral absence it should not be done until maturity has been reached and only at that time if necessary for functional or cosmetic reasons

tomatic or may be painful as in the case of the tarsal navicular and the tibial tubercle These osseous structures are sites of predilection for aseptic necrosis The accessory navicular or divided navicular has also been termed extranavicular, os tibiale externum or prehallux The existence of this condition has been known to anatomists for many years It was only after the widespread utilization of the roentgen method of examination that a thorough investigation of this anomaly was made It is probably present in 4 to 5 per cent of all persons and not infrequently is associated with pain and tenderness The pain persists despite the use of plaster casts and other conservative methods of treatment It is often mistaken for a fracture of the tuberosity of the navicular Dwight in 1907 divided it into two types (1) the true tibiale externum, which should be considered a part of the skeleton and represents the isolated tubercle of the navicular, and (2) the false tibiale externum, a small rudimentary ossicle farther out in the tendon of the tibialis posticus muscle This supernumerary bone is present in many lower animals to a greater or lesser degree of development and may have considerable functional importance It has been suggested by Monahan that it is a direct evolutionary descendent of the prehallux or sixth toe which occurs in vertebrates such as the opossum

The accessory navicular develops as a preformed cartilage in the second month of fetal life It does not manifest itself by roentgen methods until the age of nine or in many cases eleven years The accessory navicular is usually well formed and of uniform density It represents the largest regular anomaly of the tarsals The true tibiale externum develops as an ossification center of the navicular and instead of uniting normally with the body of the bone continues its independent development to form an extra ossicle It represents a portion of the navicular bone which envelops the internal aspect of the head of the talus and is separated from the navicular itself by a transverse fissure It is situated at the insertion of the tendon of the tibialis posticus muscle and is retained firmly in position by the tendon

The tendon of the tibialis posticus divides into two chief divisions, a deep and a superficial The deep portion becomes attached principally to the tubercle of the navicular and in part to the first cuneiform When the accessory navicular is present, the tendon attaches to it instead of to the tubercle of the navicular The smaller pea sized rudimentary tibiale externum is the one commonly seen and is situated more laterally in the tendon of the tibialis posticus and in most instances is separated from the navicular itself The true tibiale externum is usually responsible for pain tenderness, and prominence along the mesial portion of the foot It performs some of the duties of the usual one piece navicular It is usually bilaterally symmetrical although the size may vary in the two feet There may be a distinct ossicle separate from the main bone on one side while on the other side there is an enlarged or hooked appearance due to the fact that the two elements are united

The separate ossicles may cause pain for several reasons There may be a traumatic synovitis of the posterior tibial tendon, the anomalous bone causing pressure and irritation of the tendon sheath The presence of an accessory navicular of moderate or large size results in displacement of the tendon of the tibialis posticus and its direction of pull is changed This results in faulty mechanics of the foot with pain on use Bursitis is common in this region and is due to mechanical irritation Secondary inflam

this bone, the articular cartilage of the metatarsal being prolonged for this purpose The two sesamoids are separated from each other by the tendon of the flexor hallucis longus Other sites of sesamoids in the feet are in the region of the metatarsophalangeal joints the interphalangeal articulations, the lumbricals and the interossei A thick fibrous pad is interposed between the sesamoids and the skin In some instances the sesamoid bone is palpable in the soft tissues The reason for the presence of the sesamoids is not clearly understood It is probable that both phylogeny and functional adaptation have played an important role in the development of these bones

ACCESSORY BONES OF THE FOOT

Numerous accessory bones may occur in the foot The following are those most commonly seen

os tibiale externum, accessory navicular,
os trigonum
secondary os calcis
secondary cuboid,
intercuneiform,
uncinatum,
peroneal sesamoid, articulates with the inferior aspect of the cuboid,
intermetatarsum
paracuneiform
vesalianum,
astragalonavicular ossicle, Pirie's bone
os subtibiale, overlies the medial aspect of the distal portion of the astragalus in the region adjacent to the navicular,
ununited epiphysis of the fifth metatarsal
os talocalcaneous

The Accessory Tarsal Navicular The Os Tibiale Externum

The term accessory tarsal navicular is used to indicate an accessory element which is juxtaposed medially and posteriorly to the tubercle of the tarsal navicular In some instances it creates the appearance of an extension of the navicular rather than resembling the other accessory navicular elements such as the os supranaviculare and the os infranaviculare of the true bipartite types of navicular This anomaly is not uncommon Fusion between the accessory navicular and the true navicular may occur and may be partial or complete The accessory and the true navicular have cancellous trabecular structure and in many instances are united by a layer of soft tissue This soft tissue consists of hyaline cartilage dense fibrocartilage or a mixture of both The plate varies in thickness There is usually not a well developed freely movable joint with smooth hyaline articular cartilage capping each bone although in some instances there is a synovial lined fibrous capsule between the two bones There is frequently evidence of trauma in this region in the form of hemorrhage organizing fibrous tissue containing giant cell osteoclasts and chondroclasts Callus like repair tissue may be located subchondrally

There are throughout the body many primary and secondary centers of ossification which may remain unfused The unfused centers may be asymp-

tomatic or may be painful as in the case of the tarsal navicular and the tibial tubercle These osseous structures are sites of predilection for aseptic necrosis The accessory navicular or divided navicular has also been termed extranavicular, os tibiale externum or prehallux The existence of this condition has been known to anatomists for many years It was only after the widespread utilization of the roentgen method of examination that a thorough investigation of this anomaly was made It is probably present in 4 to 5 per cent of all persons and not infrequently is associated with pain and tenderness The pain persists despite the use of plaster casts and other conservative methods of treatment It is often mistaken for a fracture of the tuberosity of the navicular Dwight in 1907 divided it into two types (1) the true tibiale externum, which should be considered a part of the skeleton and represents the isolated tubercle of the navicular, and (2) the false tibiale externum, a small rudimentary ossicle farther out in the tendon of the tibialis posticus muscle This supernumerary bone is present in many lower animals to a greater or lesser degree of development and may have considerable functional importance It has been suggested by Monahan that it is a direct evolutionary descendent of the prehallux or sixth toe which occurs in vertebrates such as the opossum

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matory changes in the surrounding soft tissues are frequent The extra bone bears much of the weight of the foot and is a constant point of pressure with resultant swelling and irritation of the soft tissues The condition is characterized by pain in the region of the tubercle of the navicular There is usually marked tenderness and there may also be redness and swelling

In differential diagnosis several conditions must be considered The accessory bone is often mistaken for a fracture, the presence of a prominence on the inner side of the foot with pain and tenderness leading to this diagnosis A history of injury and the fact that the accessory navicular is unilateral tends to cause this error It must be stressed that an isolated fracture of the tarsal navicular is rare When fracture does occur, the fragments are sharply defined and irregular in outline After healing has taken place, one fragment may become more dense than the other because of disturbances in circulation The adjacent surfaces of the two fragments may become sclerosed The accessory navicular has a smooth contour and texture which are usually not present in fracture Comparison of the two feet is important in establishing the roentgen diagnosis In osteochondritis of the navicular, termed Kohler's disease, there is retardation of development of the bone and pain and tenderness It occurs in children between the ages of five to ten years while an inflamed or irritated accessory navicular does not appear usually before the twelfth year

Os Talocalcaneus

Hirschtick reports an anomaly which has never previously been described It consists of a hard rounded mass of bone distal and anterior to the tip of the lateral malleolus The mass appears to be fixed in some instances to the underlying bone On roentgen study there is a large osseous body on the lateral aspect of the talus and calcaneus with evidence of pressure on both of these bones The proximal end of the anomalous bone forms a shallow articulation with the talus just above and anterior to the lateral tubercle The anomalous bone curves downward and backward forming a rough semicircle and it articulates with the calcaneus just below and behind the sinus tarsi There is a concavity in the talus and a defect in the calcaneus in the region of the anomalous bone The shape of the anomalous tarsal bone resembles half of an elongated doughnut with a small hole in the center Gross and microscopic examination of the bony mass shows it to be composed of normal osseous and cartilaginous tissues Because of the location of this anomalous bone the name os talocalcaneus is suggested

PARTIAL ABSENCE OF THE BONES OF THE FEET

The structures of the peripheral joints are derived from the mesoderm During the third week of fetal life the future bones and joints become distinguishable as scleroblastoma and extend as axial rods from the vertebral column in the limb buds At about the fifth week there is condensation of the cells in these regions and the cores assume the appearance of mesenchymal links These cell structures are the progenitors of the bones The intersegmental tissues subsequently develop into the joints Failure of development of the mesenchymal cords the progenitors of the bones

results in absence of the cuneiforms, second metatarsal, phalanges, and fibula with consequent deformities and static osteoarthropathy. Condensation of mesenchymal cells in the absence of segmentation produces absence of joint formations or so called synostosis. In a case reported by Weitzner, there was congenital synostosis of the talus and navicular and also hereditary multiple ankylosing arthropathy of the hands and wrists.

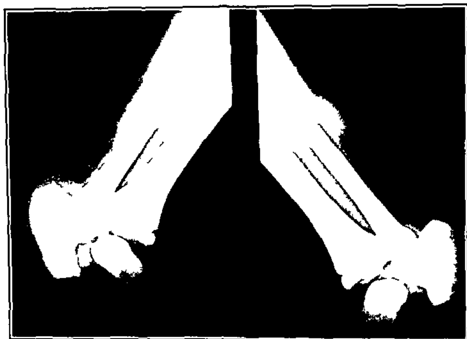


Fig 92 Congenital Malformation with Absence of the Distal Portions of the Feet. Only the talus and the calcaneus are present, the remaining bones of the tarsus and feet being absent.

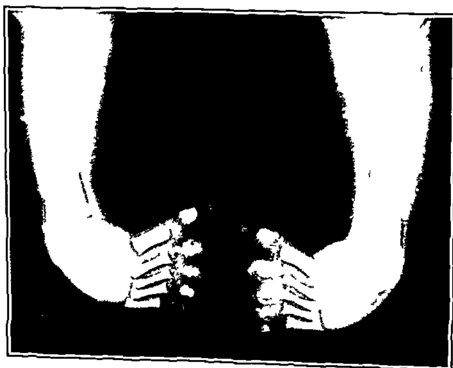


Fig 93 Club Feet. There is marked internal rotation of both feet with malposition of the bones. The bones are normally developed and ossification is proceeding at a normal rate.

CONGENITAL CLUBFOOT

The term clubfoot is used to denote a persistent, faulty supination or talipes deformity of the foot It is characterized by adduction of the forefoot, inversion, and plantar flexion as a rule The condition is usually bilateral and appears hereditary although the mode of inheritance is unknown The roentgen picture is characteristic The navicular and cuboid are displaced medialward with relation to the heads of the talus and calcaneus causing medial angulation of the anterior portion of the foot There is inward rotation of the calcaneus with relation to the talus with resultant inversion The calcaneus is displaced posteriorly, the superior surface of the bone lying in close relation to the tibia The talus projects anteriorly to the calcaneus It is widened and flattened in some instances, its proximal articular surface, which is usually smooth becomes notched The calcaneus is short and broad and its anterior aspect may be enlarged The navicular assumes a cuneiform shape The talus is displaced laterally with relation to the calcaneus and the other bones of the tarsus and the metatarsals are superimposed lying more closely in apposition to each other than normally The fifth metatarsal is hypertrophied while the first metatarsal undergoes atrophy The condition can not be diagnosed by roentgen methods in the neonatal period as the bones of the tarsus are unossified at this time As the child grows older the deformity becomes manifested clinically and roentgenographically Hypoplasia of the structures of the foot ensues and affects both the bones and soft tissues

Clubfoot is in some instances associated with retardation of muscle development This change may affect certain groups of muscles more than others Difference in the degree of maturity of muscle groups is the apparent cause of clubfoot as the more mature muscle groups exert a stronger pull The skeletal development of the foot is grossly normal and there are no differences in the sizes of the two feet

ANOMALIES OF THE SPINE

Occipital Vertebra

The condition termed occipital vertebra is the result of incomplete assimilation of the most posterior of the three scleromeres which form the base of the skull In consequence the area about the foramen magnum develops characteristics similar to a vertebral segment There is a hyper chordal arch which is partially or completely fused to the anterior margin of the foramen magnum In rare instances this bears an articular surface for the third condyle A partial or complete neural arch is outlined above the dorsal surface of the foramen The transverse processes may be absent When they are present they may be fused to the base of the skull and do not have foramens for the vertebral artery The masses which bear the condyles may encroach upon the foramen magnum with resultant deformity of the foramen The condyles are normal in appearance The so called third condyle may be present in the anterior part of the foramen as a separate ossicle The condition known as occipital vertebra must not be confused with atlanto occipital fusion

Atlanto-occipital Fusion

Since any level of the spine may under certain conditions assume the characteristics of the level immediately above or below, one of these segments may assume the vertebral form, or a complete or partial fusion of the atlas and occiput may result The ring of the atlas may be incomplete posteriorly In some instances it is fused to the occiput unilaterally or bilaterally Roentgenograms with the head in flexion and extension show that it does not separate from the base of the skull upon marked forward flexion There may be fusion of two or more cervical segments Flattening of this area comprises platybasia

Basilar Impression Platybasia

Basilar impression is characterized by elevation of the first cervical segment so that it extends above a line drawn from the hard palate to the posterior margin of the foramen magnum the so called Chamberlain line Platybasia occurs in two forms The congenital type with associated atlanto occipital fusion is characterized by flattening of the occipital bone, distortion of the foramen magnum and decrease in its antero posterior diameter The acquired type is secondary to a softening of the base of the skull and occurs in Paget s disease, osteomalacia osteogenesis imperfecta, lipoidosis hyperparathyroidism, cleidocranial dysostosis, senile osteoporosis, and rickets The skull is invaginated by the cervical spine and the base of the skull is thrust upward into the posterior fossa The basilar portion of the skull may become convex upward presenting a reverse curvature The foramen magnum lies at the level of the petrous pyramids and is funnel shaped The base of the skull may develop a recess to accommodate the atlas There is no fusion between the atlas and the occiput, studies in flexion and extension demonstrating normal range of movement between the base of the skull and the atlas The petrous pyramids are elevated and distorted The clivus lies on the same plane as the floor of the anterior fossa The neck is short and its movements are decreased The head may be held at an angle There is apt to be exaggeration of the normal anterior cervical curvature and the foramens appear smaller than normal There may be resultant compression of the cord or cranial nerves Compression of the cerebellum may force a part of its substance to herniate downward into the cervical canal, the so called Arnold Chiari malformation

Neurological symptoms occur and are often erroneously ascribed to multiple sclerosis, spastic paralysis amyotrophic lateral sclerosis, cerebellar or upper cervical canal tumor, Klippel Feil syndrome, hydrocephalus, syringomyelia and other neurological disorders Since decompression of the foramen magnum and upper cervical region may afford hope of arresting the destructive process in the central nervous system, lateral survey roentgenography of the upper cervical spine in full forward flexion is indicated in all patients with neurological manifestations of upper cord degeneration or compression Atlanto occipital fusion, occipital vertebra, and ossiculum terminale usually cause symptoms by constriction of the foramen magnum Basilar impression is an acquired distortion and causes pressure within the cerebellar fossa by invagination of that segment Early operation offers promise of arresting the progress of the condition although restoration of the normal relationship may be impossible

Agenesis of the Dens

Congenital absence of the dens is an uncommon lesion It may cause no symptoms and in some instances the existence of this condition is discovered only during a roentgen examination after an injury Many of the patients with this anomaly lead a perfectly normal life The diagnosis is established only by roentgen study In differential diagnosis it is necessary to consider the possibility of a fracture during the early years of life with the subsequent atrophy or absorption of the dens



Fig 94 Partial Absence of the Dens *A* Anteroposterior roentgenogram The dens is shortened and incompletely developed *B* Lateral laminogram The anomaly of the dens is clearly illustrated The routine lateral roentgenogram of the cervical spine did not show the anomaly clearly

The Klippel-Feil Syndrome Congenital Fusion of the Cervical Vertebræ Brevicollis

Introduction The Klippel Feil syndrome is a term used extensively in the literature to denote a clinical condition characterized by congenital fusion of two or more cervical vertebræ There are usually other associated congenital anomalies of the spine and other portions of the skeleton and neurological disturbances It is important in the differential diagnosis of pain or deformity of the neck and in the analysis of neurological symptoms referable to the cervical segments of the spinal cord

Hutchinson in 1894 and Clarke in 1906 reported cases of the syndrome However the condition is generally named after Klippel and Feil as they were the first to record in 1912 a complete clinical description with subsequent postmortem studies They termed the lesion a ' cervical thorax In 1919 Feil collected and reviewed all the previously reported cases and outlined the characteristics of the syndrome He divided the condition into three types (1) those with complete fusion of the cervical spine (2) cases of partial coalescence of the cervical segments and (3) those with reduction of the number of cervical vertebræ and fusions and anomalies



Fig 95 Klippel Feil Syndrome There are multiple congenital anomalies and coalescence of the first second third fourth and fifth cervical vertebræ The cervical lordosis is markedly increased There is platybasia the odontoid process rising to a higher level than normally and elements *of the cervical spine lying above* Chamberlain s line consistent with basilar impression

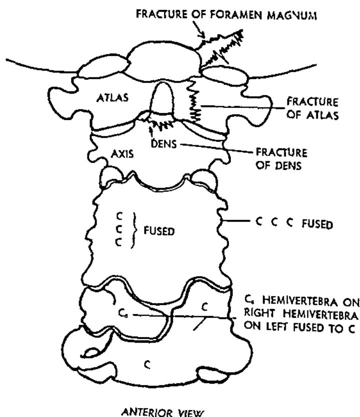


Fig 96 Klippel Feil Syndrome Schematic diagram anterior view of cervical vertebræ as found at *postmortem examination* There are fractures of the foramen magnum atlas and dens The third fourth and fifth cervical vertebræ are fused The sixth cervical vertebra is in the form of two hemivertebræ that on the left being *fused to the* seventh cervical vertebra

in other parts of the spinal column Feil's essential clinical manifestations comprised shortness of the neck, low implantation of the hair line, and limitation of the motions of the head and neck The secondary characteristics which he did not find in every instance, consisted of scoliosis or kyphoscoliosis, elevation of the scapula descent of the nipples, torticollis of muscular or osseous origin, and disproportion of the length of the extremities which resulted in a simian like appearance

Etiology and Embryology The most generally accepted opinion is that the syndrome is the result of a widespread failure of normal segmentation of the mesodermal somites In the third week of development of the fetus, paired masses of mesoderm on each side of the closing neural tube become divided progressively into paired lateral segments, the so called mesodermal somites The cells of each somite become arranged about a central canal, the myocoele The ventromedial wall of the somites, termed the sclerotome, breaks down and its cells together with those in the myocoele stream toward and surround the notochord These cells tend to cluster around the notochord, the cells from each group being formed from two adjacent pairs of somites The intersegmental clusters consolidate and from them extend paired lateral and dorsal processes which become respectively the transverse processes and the neural arches Chondrification centers develop to form a cartilaginous vertebra, and in the seventh week of fetal life ossification centers form, one, or according to some authors two, for the centrum one for each neural arch and one for each costal process The central and neural ossification centers meet and fuse in the fourth or fifth year of life The determination of the exact etiology of the Klippel-Feil syndrome and its related defects must await further research into the abnormal fusions and migrations of sclerotomal cells during the third to seventh weeks of development of the fetus Ingalls reduced the atmospheric pressure to which 300 pregnant mice were subjected during the early days of pregnancy and produced various specific bony congenital defects in the 2,500 embryos Certain skeletal anomalies were produced by subjecting the mother mouse to anoxia on the eighth day of pregnancy The congenital elevation of the scapula is apparently secondary to the primary vertebral lesions as elongation of the neck and descent of the scapula do not occur before the ninth week of development of the embryo The embryology of the vertebræ and the intervertebral discs has been well described by Beadle and Jordan and Kindred Mercer in 1950 demonstrated indentations of the anterior aspects of the vertebral bodies which he believed were due to delayed fusions of the primitive notochordal segments

Pathology Postmortem studies have been performed on less than ten of the reported cases The original case of Klippel and Feil died of pneumonia pleuritis and nephritis all of which were unrelated to the syndrome Jarcho and Levin describe normal spinal cord sections in their case Avery and Rentfro recorded the case of an infant that died thirty five days after birth of inanition and pneumonia At postmortem there was found a cleft involving the occipital bone and the cervical and first four dorsal neural arches with enlargement of the foramen magnum and prolapse of the medulla and cerebellum Microscopic sections showed a division of the upper part of the spinal cord by a deep anterior fissure doubling of the lower portion of the canal and a moderate degree of gliosis

Clinical Manifestations The Klippel Feil syndrome comprises primarily a fusion of two or more cervical vertebræ and manifests itself

clinically by a shortening and limitation of mobility of the neck. In association with the malformations in the cervical region, there are in many instances anomalies of other segments of the spine, congenital abnormalities of other portions of the skeleton and neurological disturbances. If the vertebral coalescence is slight in degree and other associated spinal anomalies are absent, the condition may produce no clinical manifestations and go unrecognized unless roentgen studies are performed. In the more marked cases, the patient presents a characteristic clinical picture and the diagnosis can often be established definitely by inspection alone. The neck is markedly foreshortened or entirely absent. The head is sunken and implanted on the chest. The hair line posteriorly is very low, extending to the upper limit of the thorax. The head is, in many instances, maintained in a position of torticollis. Other less common manifestations comprise facial asymmetry, difficulty in breathing and swallowing, and shortness of breath. The movements of the head are usually limited although there is freedom from pain on motion. Meningocele in the cervicodorsal region rarely accompanies the syndrome. Syringomyelia, radiculitis of the cervical roots due to constriction and narrowing of the foramina, platybasia, and defects in the foramen magnum such as coarctation and asymmetry, are rare concomitants of the syndrome.

It is important to distinguish congenital fusions of the cervical spine from tuberculosis of the spine, congenital torticollis, old healed fractures and other similar conditions in which therapy offers hope of improvement or alleviation of symptoms. It is in most instances not advisable to utilize extensive or formidable modes of therapy, efforts being confined to symptomatic and supportive measures. Individuals with the Klippel-Feil syndrome are particularly prone to fractures of the cervical spine because of the limited or absent mobility of the neck and they should therefore refrain from diving and arduous sports such as wrestling or football in which there is violent bodily contact. Traumatism to the head or neck should be avoided. The prognosis is good and as a rule the condition is compatible with a long life. The associated muscular and orthopedic deformities or the combined congenital and neurological lesions may alter the prognosis and require special forms of therapy.

Roentgen Manifestations. The roentgen examination is essential to establish the diagnosis and to determine the character and extent of the malformations in the cervical spine as well as other skeletal anomalies. Two or more of the vertebral masses are solidly united into a single bony mass with complete absence of the intervertebral spaces and discs. The lateral masses are irregular or fused and the spinous processes are small, deficient or bifid. The intervertebral foramina are smooth in contour but are frequently smaller than normal and oval rather than circular in shape. Fusion of the atlas and occiput is common. Anomalies of the more caudal portions of the spine comprising anterior and posterior spina bifida, sacralization of the last lumbar vertebra and hemivertebræ may be found. Scoliosis and kyphosis are occasionally present. Roentgen studies of the skull should always be performed in order to demonstrate associated deformities such as platybasia and coarction, asymmetry and other malformations of the foramen magnum. Congenital elevation of the scapula and cervical ribs are often present, the extent of these changes varying considerably on the two sides. There is elevation of the apices of the lungs.

The technical aspects of the roentgen examination are of the greatest importance to demonstrate the character and extent of the changes. Be-

cause of the short neck and sunken head, roentgenograms of perfect detail may be obtained only with considerable difficulty This is particularly true of the anteroposterior and oblique projections as the occiput and mandible overlie and obscure the cervical vertebræ Roentgenograms with the jaw in rapid motion may be helpful in making the anteroposterior films Laminography may be essential in cases with extensive coalescence of the cervical vertebræ In infants and young children laminagrams are unsatisfactory due to lack of contrast Stereoscopic



Fig 97



Fig 98

Fig 97 Klippel Feil Syndrome Lateral view of cervical spine The first cervical vertebra is elongated and tilted The second to sixth cervical vertebræ are anomalous and fused The normal cervical lordosis is completely absent

Fig 98 Spina Bifida of the Cervical and Dorsal Spine There is extensive spina bifida involving the lower cervical and upper dorsal vertebræ

roentgenograms are essential and should be utilized in every instance The roentgen studies should include not only the cervical region but also the remainder of the spine and other parts of the body as indicated

The Klippel Feil syndrome was in the past considered a rare and unusual condition It is much more common than previously supposed and must be considered in the differential diagnosis of all malformations of the upper spine With the greatly increased application of roentgen methods of examination in present day practice many new cases will be *found and recorded* and the syndrome will take its rightful *place in* clinical practice as a relatively frequent lesion

Spina Bifida

Failure of the neural arch to fuse posteriorly is the most common congenital anomaly of the spine The laminæ are absent partially or completely and the spinous processes may also be absent The term spina bifida occulta is used to describe those instances in which the defect involves only the bony structure One or more vertebræ may be affected The fifth lumbar vertebra and upper sacrum are the commonest sites of this lesion although any portion of the spine may be involved If the condition is accompanied by a meningocele, it is termed spina bifida vera

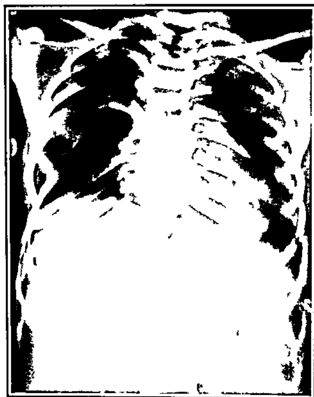


FIG 99 Hemivertebra with Anomalies of the Ribs and Scoliosis There is a hemivertebra on the left between the seventh and eighth dorsal vertebræ with anomalies of the ribs on the right side and a marked degree of scoliosis to the right

Hemivertebra

Defects in the body of the vertebræ are usually due to lack of growth of one of the ossification centers Asymmetry in the growth of the centers may result in the formation of a hemivertebra One or several hemivertebræ may be present In the dorsal region there are usually variations in the ribs such as absence of the rib on the side of the hemivertebra as the embryonic development of the vertebral bodies and the corresponding ribs is closely associated

Supernumerary Vertebræ

This anomaly is common Any portion of the spine may be involved The lumbar spine is the commonest site of this condition and the presence of six lumbar vertebræ is frequent If the extra vertebra is normal, no symptoms or deformity result Partially formed or wedged segments

result in marked spinal curvature Studies of the entire spinal column are essential to establish the correct diagnosis Variations in the number of the dorsal and lumbar vertebræ are due to aberrations in segmentation during fetal life Occasionally an individual has only eleven ribs and a supernumerary vertebra may be diagnosed erroneously Usually the extra vertebra is normal in size and shape and causes no symptoms Variations in the number of sacral segments may compensate for an abnormal number of lumbar vertebræ

Intrathoracic Meningocele

Intrathoracic meningocele is extremely rare Pohl is credited with having reported the first case in 1933 In his patient, the meningocele arose from the intervertebral foramen at the fourth dorsal vertebra and projected laterally at the apex of a well marked kyphoscoliosis There were erosions of the posterior aspects of the ribs and widening of the intervertebral foramen Until 1952 only 15 cases had been recorded in the literature The condition is commonly associated with kyphoscoliosis and is characterized by the presence of a posterior mediastinal mass projecting laterally through the intervertebral foramen with enlargement of the foramen and absorption of the adjacent portions of the ribs There are frequently associated anomalies such as bifid vertebræ In a large percentage of the cases the condition is associated with Recklinghausen s disease The presence of a posterior or paravertebral mass enlargement of the foramens and absorption of the adjacent ribs should suggest the possibility of a meningocele Masses which show evidence of increase in size usually require surgical excision with closure of the dural sac The size varies from 3 to 4 cm in diameter to a mass which occupies the entire thoracic cavity on one side On myelographic studies the opaque material enters the thoracic dural sac in small amounts The intervertebral foramens in the region of the mass are enlarged and there is kyphoscoliosis at the point of origin The lesion is usually asymptomatic This is in striking contrast to neurofibromas which commonly produce neurological symptoms The absence of neurological manifestations is explained by the fact that the dural herniation does not enclose the spinal cord and the nerve tissues remain intact

Intrathoracic Meningocele with Spinal Deformity and Multiple Neurofibromatosis Multiple neurofibromatosis is characterized by protean pathological changes Approximately one case in ten shows skeletal changes and in 50 per cent of these the spine is involved There is an apparent association of involvement of the spine with the very rare condition termed lateral intrathoracic meningocele Up to the year 1949 only 5 such cases had been reported Kessel in 1951 reviewed reports of 8 cases of lateral intrathoracic meningocele from the literature and described two new cases Seven of the cases showed an association with multiple neurofibromatosis indicating that there is relationship between the two diseases Roentgen examination showed marked spreading of the sixth seventh and eighth ribs with erosions of the margins of these ribs The pedicles of the fifth sixth, and seventh thoracic vertebræ were absent and there was scalloping of the posterior margins and anterior wedging of the affected vertebræ with preservation of the intervening disc spaces A roentgenogram of the chest showed a soft tissue opacity extending into the lung field in the region of the above described change An acute kyphoscoliosis was pres

ent at the level of the lesion The roentgen appearances were those of localized bone resorption from the pressure of a tumor mass Myelography may reveal an obstruction at the level of the lesion

Block Vertebra Congenital Fusion of the Vertebræ

Block vertebra is characterized by poor segmentation with fusion of the bodies of two or more vertebræ into a single bony mass Evidence of the intervertebral disc usually remains Block vertebræ may occur in any portion of the spine



FIG 100 Congenital Fusion of the Third and Fourth Lumbar Vertebræ *A* Anteroposterior view *B* Lateral projection There is absence of the intervertebral space between the third and fourth lumbar vertebra with congenital fusion of the vertebræ The condition easily may be confused with tuberculosis or other disease of the spine

Dilatation of Vertebral Canal Associated with Congenital Anomalies of the Spinal Cord

The spinal canal may become markedly dilated in the presence of an intraspinal neoplasm Equally pronounced dilatation may occur as the result of a congenital anomaly of the cord structures The enlargement of the spinal canal occurs early in the development of the embryo, probably at the time the ossification centers are laid down, as there is no evidence of spinal cord compression on lumbar puncture or at operation At the third month of intrauterine life the spinal cord segments correspond anatomically to the vertebral segments At birth, the lower end of the spinal cord is at the level of the third lumbar vertebra and in adult life at the first lumbar vertebra In the lumbar region of the spinal cord, the usual site of myelodysplasia failure of upward migration causes tension on the spinal cord and accounts for the development of symptoms early in adult life Enlargement of the spinal canal has been described

in certain instances of diastematomyelia In some cases of spina bifida occulta, there is dilatation of the vertebral canal The dilatation of the canal associated with congenital anomalies cannot be distinguished from that due to intraspinal neoplasm as the pedicles are thinned in both instances Walker reports three cases of myelodysplasia associated with a dilatation of the spinal canal at the site of the anomaly In one instance, there was a vascular abnormality an angioma and aneurysm of the spinal cord accompanied by enlargement of the spinal canal Splitting of the spinal cord or diastematomyelia is very rare in the cervical region The majority of the cases occur below the mid thoracic level



FIG 101 Spina Bifida Widening of the Canal Myelodysplasia There is marked widening of the spinal canal in the lower dorsal and lumbar regions The bony septum within the spinal canal and the anomalies of the vertebral bodies are clearly visualized

Diastematomyelia

Diastematomyelia is a congenital malformation of the neural axis characterized by an abnormal cleft or division of the spinal cord or its derivatives occurring in association with developmental anomalies of the spine such as spina bifida The division of the cord or cauda equina is sagittal and the two segments are separated from each other by a septum of bone or fibrocartilage The septum is attached anteriorly to one or more vertebral bodies and posteriorly to the dura In some instances the posterior attachment extends to the malformed arches of the affected vertebræ The septum of osseous or fibrocartilaginous tissue which transfixes the cord or the cauda equina fixes the cord in a low anatomical position and results in impairment of its ascent during the normal growth

of the spine Adhesions frequently occur in the region of the septum Traction on the cord or cauda equina results in neurologic manifestations which increase progressively as the patient grows The lesion is most common in the lumbar region but has also been encountered in the middle and lower thoracic regions

The condition is important as it is usually associated with neurologic disturbances affecting the lower extremities or interference with sphincteric control At birth, an abnormal tuft of hair, a skin dimple, a localized accumulation of subcutaneous fat, a port wine stain, a nevus formation or other cutaneous lesion suggestive of the presence of spina bifida may be present Meningocele may occur in association with diastematomyelia In some instances, there are no clinical manifestations, careful questioning being necessary to establish the presence of slight alterations

The infant as a rule reveals no abnormalities of the lower extremities at birth However, the child either fails to walk correctly at the usually expected time or has a limp early in life, abnormality of the gait being the manifestation which first calls attention to the condition There may be muscular imbalance of the lower extremities On physical examination there is atrophy of one or both legs One of the feet may be shortened or present a deformity such as talipes, metatarsus varus, or pes cavus The deep reflexes of the lower extremities are hyperactive, diminished or in some instances absent Ankle clonus and dorsiflexion to the Babinski stimulus may be elicited Hyperesthesia is often present in the legs and urinary and fecal incontinence is frequently seen Increasing eneuris is common The hypothesis has been advanced that diastematomyelia is due to an incipient form of twinning, the duplication of the cord being the only well developed persisting duplication This theory explains the presence of complete meninges and complete sets of nerve tracts in each of the two tubes at the level of the deformity The alteration of the cord is usually limited to a small segment and is associated with bone changes in most instances

Roentgen Manifestations The criteria for diagnosis are definite The spine at the level of the lesion shows widening of the neural canal and the interpedicular spaces These changes may involve as many as six adjacent segments of the spine The affected pedicles usually are not narrowed This is of particular significance as it indicates the presence of widening of the spinal canal on a developmental basis and is important in differentiating the condition from the erosion associated with intraspinal tumors which produce erosion of the pedicles The septum dividing the cord is demonstrable only if it is osseous, in which instances it is visualized on the sagittal roentgenogram as an irregular linear area of bony density in the midline in the neural canal The bony septum measures 1 0 to 1 5 cm or less in length While not usually demonstrable in the lateral projection, the spicule of bone in some instances may be seen in the region of the posterior surface of the vertebral body or the posterior portion of the neural canal in relation to the lamina

Associated anomalies of the bodies of the vertebræ are of relatively frequent occurrence There may be a decrease in the anteroposterior diameter of the vertebral bodies Double hemivertebræ may be present One or more of the bodies may be hypoplastic or unsegmented and the intervertebral spaces may be narrowed This results in kyphosis and scoliosis Anomalies of the vertebral arches with overgrowth posteriorly are common There is often fusion of the laminæ with diagonal fusions

of the contralateral halves or fusion of adjacent arches on one side Absence of fusion of the arch with spina bifida is usually seen Myelography is not necessary to establish the diagnosis in cases in which the bony spicule is visualized within the widened neural canal, although myelographic studies may afford valuable data in certain doubtful cases The septum is manifested by a rounded or elongated, irregular filling defect in the central portion of the opaque column, the bony spicule lying within the defect Associated fibrous or fibrocartilaginous tissue increases the size of the defect Arachnoidal adhesions result in irregularity of outline and deformation of the opaque column The presence of a non opaque septum can be proven only by myelography prior to operation or autopsy As the age of the patient increases the diagnosis of diastematomyelia becomes increasingly difficult This applies to both the plain roentgenograms and myelography because the concomitant abnormalities result in shortening and the development of increasing degrees of kyphosis and scoliosis In late childhood or during adult life roentgen manifestations are obscured or become impossible of demonstration

Differential Diagnosis In arachnoiditis, myelography may reveal filling defects and irregularities of outline in the opaque column The absence of widening of the spinal canal at the site of the lesion serves to differentiate the condition from diastematomyelia Tumors of the expanding intraspinal type are usually associated with narrowings of the pedicles and erosions of the posterior aspects of the vertebral bodies Intraspinal meningocele and lipoma produce widening of the neural canal but do not present a midline osseous spur or fibrocartilaginous septum The developmental anomalies of the vertebrae associated with diastematomyelia may also occur with intraspinal dermoids lipomas and meningoceles, but there is no septum in any of these lesions The bony spur occurs in only about one fourth of the cases As with other rare and unusual conditions the diagnosis will be established only if the condition is borne in mind and the roentgen as well as the clinical manifestations are clearly understood Myelography is helpful, particularly in those with a fibrous or fibrocartilaginous septum and no bony component

Therapy It is essential that the midline septum be removed in order to prevent symptoms or arrest of development Otherwise the fixation and compression of the cord or the corda equina become increasingly more marked and complications ensue Progressive neurologic impairment of the lower extremities and the rectal and bladder sphincters may result from increasing distortion of the neural axis These disturbances are the result of fixation of the spinal cord produced by the bony spicule during the growth period Surgical treatment is advised as a prophylactic measure rather than as a curative one Laminectomy with extradural removal of the bony spicule insofar as possible is essential This is followed by opening of the dura with excision of its reflections adjacent to the spicules removal of the remaining bony prominence to the anterior dura and division of all adhesions involving the bifid cord or cauda equina

ADDITIONAL READING

MATSON D D WOOD R P CAMPBELL J B and INGRAHAM F D Diastematomyelia Congenital Cleft of the Spinal Cord Diagnosis and Surgical Treatment Ped *6* 98 1950

MAXWELL H P and BUCY P C Diastematomyelia J Neurol and Exp Neur *5* 165 1946

NEUHAUSER E B D Diastematomyelia Radiology *54* 659 1950

Lumbosacral Anomalies

Anomalies in the lumbosacral portion of the spine are common and occur in a relatively high percentage of patients with and without symptoms referable to the lower portion of the back The commonest developmental or congenital anomalies are asymmetrical lumbosacral facets and sacralization of the fifth lumbar vertebra Less frequent anomalies in this region are spina bifida occulta (Fig 102), lumbarization of the first



Fig 102 Spina Bifida There is a marked degree of spina bifida involving the *fourth* and fifth lumbar vertebræ and the sacrum

sacral segment, spondylolisthesis impinging spinous processes, incomplete neural arch, and accessory articular processes In a small percentage of the cases, two or more of these anomalies may be present simultaneously In patients with backache, there is a slightly higher percentage of combined anomalies There is no definite relationship between anomalies in the lumbosacral region and the presence of radiating pain, nor between narrow lumbosacral discs and lumbosacral anomalies, although these conditions also may coexist

of the contralateral halves or fusion of adjacent arches on one side. Absence of fusion of the arch with spina bifida is usually seen. Myelography is not necessary to establish the diagnosis in cases in which the bony spicule is visualized within the widened neural canal, although myelographic studies may afford valuable data in certain doubtful cases. The septum is manifested by a rounded or elongated, irregular filling defect in the central portion of the opaque column, the bony spicule lying within the defect. *Associated fibrous or fibrocartilaginous tissue increases the* size of the defect. Arachnoidal adhesions result in irregularity of outline and deformation of the opaque column. The presence of a non-opaque septum can be proven only by myelography prior to operation or autopsy. As the age of the patient increases the diagnosis of diastematomyelia becomes increasingly difficult. This applies to both the plain roentgenograms and myelography because the concomitant abnormalities result in shortening and the development of increasing degrees of kyphosis and scoliosis. In late childhood or during adult life roentgen manifestations are obscured or become impossible of demonstration.

Differential Diagnosis. In arachnoiditis, myelography may reveal filling defects and irregularities of outline in the opaque column. The absence of widening of the spinal canal at the site of the lesion serves to differentiate the condition from diastematomyelia. Tumors of the expanding intraspinal type are usually associated with narrowings of the pedicles and erosions of the posterior aspects of the vertebral bodies. Intraspinal meningocele and lipoma produce widening of the neural canal but do not present a midline osseous spur or fibrocartilaginous septum. The developmental anomalies of the vertebræ associated with diastematomyelia may also occur with intraspinal dermoids, lipomas and meningoceles, but there is no septum in any of these lesions. The bony spur occurs in only about one fourth of the cases. As with other rare and unusual conditions, the diagnosis will be established only if the condition is borne in mind and the roentgen as well as the clinical manifestations are clearly understood. Myelography is helpful particularly in those with a fibrous or fibrocartilaginous septum and no bony component.

Therapy. It is essential that the midline septum be removed in order to prevent symptoms or arrest of development. Otherwise, the fixation and compression of the cord or the corda equina become increasingly more marked and complications ensue. Progressive neurologic impairment of the lower extremities and the rectal and bladder sphincters may result from increasing distortion of the neural axis. These disturbances are the result of fixation of the spinal cord produced by the bony spicule during the growth period. Surgical treatment is advised as a prophylactic measure rather than as a curative one. Laminectomy with extradural removal of the bony spicule insofar as possible is essential. This is followed by opening of the dura with excision of its reflections adjacent to the spicules, removal of the remaining bony prominence to the anterior dura, and division of all adhesions involving the bifid cord or cauda equina.

ADDITIONAL READING

MATSON, D. D., WOOD, R. P., CAMPBELL, J. B., and INGRAHAM, F. D.: Diastematomyelia: Congenital Cleft of the Spinal Cord; Diagnosis and Surgical Treatment. Ped. *6*: 98, 1950.
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Fig. 102. Spina Bifida. There is a marked degree of spina bifida involving the fourth and fifth lumbar vertebræ and the sacrum.

sacral segment, spondylolisthesis, impinging spinous processes, incomplete neural arch, and accessory articular processes. In a small percentage of the cases, two or more of these anomalies may be present simultaneously. In patients with backache there is a slightly higher percentage of combined anomalies. There is no definite relationship between anomalies in the lumbosacral region and the presence of radiating pain, nor between narrow lumbosacral discs and lumbosacral anomalies, although these conditions also may coexist.

Spondylolisthesis

The term spondylolisthesis is derived from the Greek "spondylus," a vertebra, and "olisthesis," meaning a slipping, and is used to designate the condition characterized by anterior displacement of a vertebra with relation to the adjacent caudad vertebra or the first sacral segment. The condition usually occurs in the lower lumbar region. Spondylolysis constitutes a defect in the interarticular portion of the neural arch without associated vertebral displacement. Spondylolisthesis comprises anterior displacement of the vertebral body and its superior articular processes with relation to the inferior articular processes and the spinous process and occurs only in the presence of a defect in the interarticular portions of the neural arch. The incidence of spondylolisthesis is estimated to be about 5 per cent. The lesion may be traumatic or congenital in origin. The transition of spondylolysis to spondylolisthesis has been demonstrated in many instances.

The relative significance of trauma and developmental defects in spondylolisthesis is still undetermined. Spondylolysis is a well recognized clinical entity. Spondylolisthesis is considered an abiotrophic disorder in which the abnormal disposition but not the definitive manifestation is present at birth. The abnormality usually develops during youth or in adult life. The vertebral arch develops by perichondral ossification rather than from a bony center. Spondylolisthesis may occur in association with abnormalities of the articular facets, elongation and narrowing of the isthmus or bony discontinuity of the isthmus, manifestations *of faulty development. It does not occur in infants and young children.* The age incidence does not increase after the age of ten to twenty years. In many cases the disease is present without symptoms and is overlooked. Spondylolisthesis does not develop in individuals with an intact isthmus.

Roache and Row in a recent study of four thousand two hundred skeletons found the incidence of defects of the neural arch or spondylolisthesis to be 4.2 per cent. In this series the fourth and fifth lumbar vertebrae were affected in 94.6 per cent. The incidence was greater in the white male than in the Negro male and almost twice as great in the white female as in the Negro female. The lesions are found with approximately equal frequency in all age groups from twenty to eighty years. Hubeny in 1931 recommended the use of oblique projections to show the extent of the defect in the neural arches. The importance of this procedure has not been widely appreciated and it has not been used to its fullest advantage. The lesion has by many authorities been termed a congenital anomaly and this may be particularly important in medical legal and military cases.

Etiology. A characteristic of spondylolisthesis is the presence of a defect in the neural arch of the affected vertebra. Willis states that the defect in the pars interarticularis is due to failure in ossification similar *to that which occurs in spina bifida. He demonstrated seven areas in* which separations of the lamina may occur. In none of the instances described by him were there manifestations of fracture such as repair or resorption. Many observers favor a traumatic origin for the disease. The pars interarticularis is comprised of cartilage and a lake of blood vessels and is particularly vulnerable to fracture. The bony defect in the vertebral arch may be unilateral. In some instances trauma is significant. A

history of trauma is not present in all cases of spondylolisthesis, although it is elicited in many cases The pars interarticularis is a frail segment and a minor trauma can fracture the isthmus It is not certain whether the origin is traumatic or whether the lesion is due to a developmental defect There is evidence that trauma of minor degree can widen the defect or initiate the lesion The condition cannot be classified on the basis of present evidence as being congenital in origin in every instance

It is advantageous to classify the degree of displacement Various methods of measuring the extent of the change have been attempted A procedure suggested by Meschan appears satisfactory in the majority of cases The lateral projection is utilized, the roentgenogram having been exposed with the central beam centered over the lumbosacral junction A line is drawn between the posterior and inferior aspect of the vertebral body above the one involved and the posterior and superior lip of the vertebral body directly below the defect A second line is drawn between the posterior upper and lower lips of the superior vertebral body These lines are extended so that if they are not parallel, they intersect to form a measurable angle The position of the apex of the angle determines the degree of spondylolisthesis The angle between the two lines is measured with a protractor If, as rarely happens, the lines are parallel, the linear distance between the lines is measured Angles less than 10 degrees are classified as slight Angles from 11 to 20 degrees are considered moderate and angles greater than 20 degrees are severe Parallel lines separated by a distance of more than 3 mm are considered abnormal The method affords a means of establishing the changes produced during weight bearing, flexion, and extension In the normal, the lines intersect below or at the level of the fifth lumbar vertebra or if parallel to one another they are less than 3 mm apart In spondylolisthesis the lines practically always intersect above the slipped vertebral body



FIG 103 Spondylolisthesis There is marked anterior displacement of the fifth lumbar vertebra with relation to the first sacral segment There is a defect in the neural arch of the fifth lumbar vertebra (arrow)

Roentgen Technique A Oblique Views The right and left anteroposterior oblique views of the lumbar spine are taken by placing the patient in the recumbent position and rotating the body so that the back forms an angle of 35 to 40 degrees with the table The degree of obliquity must be varied in accordance with the degree of obliquity of the lumbar facets The facets nearer the film are parallel and horizontal and hence are most clearly visualized This is also the case with the apophyseal joints The importance of the oblique views in the demonstration of defects in the pars interarticularis cannot be overemphasized particularly in those instances in which the defect is unilateral With bilateral defects, the lateral projection may suffice The defect in the pars inter-

articularis is visualized in the oblique projection as a discontinuity with smooth or slightly serrated margins Usually, the tip of the superior articular surface of the vertebra is directed toward the defect

B *The Anteroposterior View* The anteroposterior view is helpful but is not diagnostic in every instance In advanced spondylolisthesis the body of the slipped vertebra is superimposed upon the superior border of the subjacent vertebra and its spinous process points upward giving the so called "bow and arrow" appearance Lumbosacral anomalies occur in 40 per cent of all lumbosacral examinations while the incidence of spondylolisthesis is approximately 4 6 per cent Hence this factor is not of great aid in diagnosis In some instances the anteroposterior view proves actually misleading

C *Lateral Views* In the lateral projection the defect in the pars interarticularis is visualized as a discontinuity in the neural arch at the posterior margin of the spinal canal The defect in the lateral projection usually presents rounded smooth margins The defect is visualized most clearly with the patient lying on the side and the spine flexed, and in some cases is demonstrable only in this projection In the presence of defects of the neural arch, marked beveling of the sacrum and associated hypertrophic changes occur The supero anterior margin of the sacrum is rounded and its superior surface presents a horizontal S shaped configuration The interspace below the slipped vertebra is usually narrowed There may be lipping and sclerotic changes of the bony surfaces of this interspace, the changes being due to the movement of the vertebral body on the upper surface of the sacrum The displaced vertebral body may be unstable in the presence of spondylolisthesis This is determined by comparison of roentgenograms in the recumbent and erect positions Roentgenograms in hyperextension and hyperflexion may prove of value

Posterior Displacement of Lumbar Vertebræ

Posterior displacement of a lumbar vertebra consists of backward displacement of the cephalad vertebra in relation to the adjacent caudad vertebra or the first sacral segment Willis states that the roentgen appearance of backward displacement of the fifth lumbar vertebra is merely an optical illusion due to a discrepancy between the mid sagittal diameters of the adjacent surfaces of the fifth lumbar and first sacral segments While this illusion does occur there are many instances in which there is a true subluxation based on pathological conditions and not the result of faults in technique or anatomical variations in the lumbar spine and the lumbosacral joint is a particularly common site of these variations Care is essential to establish the presence of these variations Normally the lumbar curve is lordotic The stress in the upper lumbar region is downward and backward and in the lower lumbar region downward and forward

True backward displacement may be due to degenerative processes disease trauma or congenital anomalies Most cases of retrodisplacement of the vertebræ are associated with degeneration of the intervertebral disc This results in relaxation of the longitudinal ligaments and permits of greater movement between the vertebræ and stretching of the capsules of the apophyseal joints The instability is usually associated with sclerotic changes and lipping of the articular borders of the vertebræ The disc is one of the earliest structures in the body to undergo degenerative

changes since it is subjected to frequently repeated strain and trauma Backward displacements occur most frequently in the upper lumbar region because the vertebrae in this region incline backward The lumbo sacral junction tilts forward and downward, consequently there is a greater tendency to anterior displacement in this region In backward displacement, an hour glass formation develops at the intervertebral foramens with narrowing in the ventrodorsal axis The distance between the postero inferior border of the cephalad vertebra and the anterior border of the superior articular process of the caudad vertebra is diminished The hour glass constriction must be differentiated from narrowing or encroachment on the foramens by spur formations at the postero-inferior margin of the cephalad vertebral body In some cases of posterior displacement at the lumbosacral junction, the lumbosacral angle becomes reversed The posterior portion of the fifth lumbar intervertebral disc degenerates more than the anterior portion If this is associated with a sacrum which lies horizontally, posterior displacement may occur Degeneration of the articular cartilage of the apophyseal joints also may initiate the backward displacement particularly in association with stretching of the ligaments hypermobility of the adjacent vertebrae and degeneration of the intervening discs A sudden severe injury may result in backward displacement of a vertebra or vertebrae The trauma must be severe and involve the discs, the bony structures of the spine, the apophyseal articulations or the ligaments which support the vertebrae Narrowing of the intervertebral disc occurs in association with the backward displacement A ruptured disc may be found at operation Backward displacement has been described in tuberculosis of the spine, syphilis osteomyelitis, primary and secondary bone tumors, senile osteoporosis and osteomalacia Congenital relaxation of the ligamentous structures of the spine is important Developmental factors may be responsible for early degenerative processes of the disc structures Abnormality of the architecture of the spine associated with a congenital malformation articular facets which are directed anteriorly and posteriorly in the lumbosacral region and asymmetry of the facets may produce instability of the lumbosacral articulation

Cases which roentgenographically appear to show backward displacement may be due to technical factors Faulty positioning of the patient on the roentgen table with regard to rotation and sagging of the lumbar spine, incorrect alignment of the central ray, and too short target-film distance are common causes of error Anatomical and other factors which may explain an apparent displacement are widening of the postero inferior border of the last lumbar vertebra, hypertrophic spurring of the postero inferior margin of the vertebral body, tapering of the vertebral body, sacralization of the last lumbar vertebra and flattening of the postero superior margin of the sacrum In the roentgen examination of the lumbar spine, it is insufficient to make simply an antero posterior and lateral view with the patient recumbent Various oblique projections are essential Roentgenograms with the patient in forward and backward bending as well as with right and left bending may be helpful Laminography and stereoscopic studies should be utilized in doubtful cases Distortion results from too short a target film distance and it is advisable that the roentgenograms be made with target film distance of 40 inches or more Anatomical variations of the lumbosacral junction may result in roentgen appearances which simulate backward displacement particularly if the sacrum shows a concave posterior border

Rotation and sagging of the sacrum create a false impression of retro displacement when in fact none actually exists

Melamed and Amsfield have enumerated a set of criteria on the basis of which it is possible to establish the diagnosis of posterior displacement of the lumbar vertebræ Not all of the manifestations are present at any level, the roentgen features varying somewhat from the first lumbar to the lumbosacral level In order to establish the diagnosis, the following features are essential (1) Degeneration of the interspace The roentgen manifestations of disc degeneration comprise narrowing of the intervertebral space, reactive changes at the articular margins of the vertebral segments, the vacuum phenomenon, calcification of the intervertebral disc, instability with abnormal relation of the vertebræ, an abnormal degree of mobility of the vertebræ, and alteration of the lumbar curve (2) The posterior and anterior borders of the cephalad vertebral body must be displaced posteriorly with relation to the corresponding portion of the caudad vertebra with interruption of the continuity of the lumbar curve (3) Narrowing of the intervertebral foramen in the antero posterior diameter If the intervertebral space is decreased there is also narrowing of the intervertebral foramen The anteroposterior narrowing of the foramen results in an hour-glass appearance The encroachment is due to approximation of the postero inferior margin of the cephalad vertebral body and the superior articular process of the adjacent caudad vertebra (4) Displacement of the facets and/or widening of the apophyseal joint space With facets of the internal external type widening of the overlapping articular processes in the sagittal plane is present With facets facing anteriorly and posteriorly, widening of the joint space must be present (5) Prominence or protrusion of the spinous process of the displaced vertebra in the sagittal projection (6) Alteration of the lumbar curve (7) The roentgen signs of retrodisplacement must not be eliminated by changes in the technique of the roentgen study Posterior displacement of the lumbar vertebræ has been referred to as reverse spondylolisthesis or spondylolisthesis posterior The use of the term spondylolisthesis in this regard is incorrect and the condition should be termed merely posterior displacement of the lumbar vertebra

Accessory Sacro-iliac Articulations

Accessory sacro iliac articulations are relatively frequent The antero inferior portions of the adjacent surfaces of the sacrum and the ilium form the true sacro iliac joint Only about half of the adjacent surfaces of the sacro iliac articulations are actually in contact with each other The remaining portions separated by a deep cleft one eighth to one quarter of an inch wide are connected by the posterior sacro iliac ligaments Articular facets may form a bridge across this space with the development of accessory sacro iliac joints which are entirely separate from the true sacro iliac joints These accessory articulations are diarthrodial articulations and have a joint space capsule and articular surface

Hadley describes two types of accessory sacro iliac articulations Prominent facets surmounted by articular cartilage develop at these points The accessory joints are superficial and when affected by arthritis are tender on deep pressure They are separated from the true sacro iliac joints by a foramen through which pass branches of the superior gluteal nerve and artery This foramen is a useful landmark for identify

ing the superficial accessory sacro iliac articulation It lies between the vicarious joint and the true sacro iliac joint

The accessory joints are important in that individuals with accessory articulations are prone to develop low back pain, sciatica, muscle spasm, limitation of motion and tenderness to pressure in the region of the accessory joints To establish the diagnosis the roentgen examination may be made with the patient prone or supine In some instances the joints are best visualized in the axial or tangential projection To obtain this view the patient sits at the end of the Bucky table bent forward with the central ray directed vertically downward toward the lumbosacral joint A large sandbag placed against the prominence of the sacrum lessens the fogging from scattered radiation Stereoscopic studies are helpful

Agenesis of the Lumbar Spine

Agenesis of the lumbar spine is an extremely rare anomaly, only 10 living cases having been recorded Many are stillborn Additional malformations such as imperforate anus rib fusions, Klippel Feil syndrome and partial or total absence of the genitourinary tract are common Paraplegic deformities of the lower extremities usually result In a case reported by Rosen, the first three lumbar vertebræ were represented by a synostotic mass with absence of the fourth and fifth lumbar segments The sacrum was small and incompletely developed The pelvis was narrowed and rotated Both patellas were absent There was synostosis of the bodies of the second and third cervical vertebræ, spina bifida involving the lower cervical region and the first four dorsal vertebræ, and multiple anomalies of the rib cage

Sacralization

The transverse processes of the fifth lumbar vertebra on one or both sides may be unusually wide and elongated When the transverse process articulates with the upper sacrum, the condition is termed *sacralization* This results in a potential weakness and after a trauma, patients with the anomaly suffer more and are longer in recovering than those with a normal spine The most common anomaly of the lower lumbar region is widening of one or both transverse processes of the fifth lumbar vertebra

Congenital Absence of the Sacrum and Coccyx

Congenital absence of the sacrum and coccyx is known as sacrococcygeal agenesis It is a rare anomaly and only 45 cases had been reported up to the year 1951 The condition in many instances goes undiagnosed until roentgen examination has been made although it frequently presents characteristic features which *make it easily* recognizable without roentgenograms The absence of the spinal column below the lumbar region results in obvious and marked narrowing of the pelvis and shortening of the intergluteal fold Atrophy and dimpling of the buttocks are prominent manifestations and are probably due to fibrous tissue contraction The muscles of the lower extremities show atrophy The atrophy becomes increasingly prominent as skeletal growth progresses Club foot occurs in many instances There is marked prominence of the lumbar spine

The embryological basis for the anomaly is not understood The ossification of the first sacral vertebra begins at the eighth to ninth week of fetal life and is soon followed by ossification of the second and third sacral vertebrae This indicates that the condition must develop in the early weeks of fetal life The influence of heredity must be considered Anomalies of the appendicular skeleton are inherited to a marked degree and certain axial skeletal defects are known to occur on a hereditary basis

Congenital absence of the sacrum and coccyx may vary from the absence of one coccygeal segment to complete lack of the vertebral column below the tenth thoracic segment The patients may be divided functionally into two groups (1) the first sacral segment is lumbarized or the fifth lumbar vertebra is sacralized In these instances the bony ring of



Fig 104 Absence of the Coccyx A Anteroposterior view B Lateral projection There is congenital absence of the coccyx The condition caused no symptoms and was discovered during a roentgen study of the urinary tract

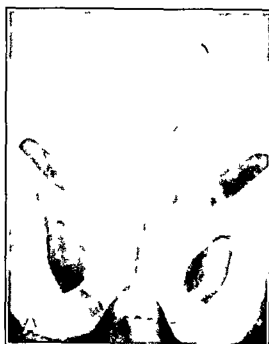
the pelvis is complete Stance and gait are normal (2) the sacrum is entirely absent and the iliac bones meet posteriorly Standing and walking are impossible and there are associated neurologic abnormalities due to interference with or agenesis of the spinal nerves Incontinence of urine and feces muscle atrophy sensory changes deformities of the lower limbs and visceral abnormalities may occur The right and left pelvic girdles are closely approximated in the midline A survey of the skeleton usually discloses no other abnormalities although in some instances there may be associated anomalies such as fusions of the ribs and bilateral dislocation of the hip In differential diagnosis it is necessary to consider anterior sacral meningocele chordoma and metastatic carcinoma

Anterior Sacral Meningocele

Anterior sacral meningocele is a congenital anomaly characterized by agenesis of the anterior portion of the sacrum protrusion of the meninges

through the bony defect, and a hernial sac in the pelvis While relatively rare, it is an important lesion which may become a serious problem in diagnosis and therapy Prior to operation or autopsy the diagnosis can be established only by roentgen methods In a recent paper by Callihan, it is stated that only approximately 52 cases had been recorded in the literature prior to 1942 The terms *occult meningocele of the sacrum and spina bifida occulta* have been used synonymously by some authors The conditions are different in that a meningocele is a cyst of the meninges or a hernia extending through the walls of the vertebral canal, while spina bifida occulta comprises a lack of junction of the laminae, with no projecting sac An occult meningocele is a diffuse dilatation of the dural sac with or without the associated element of herniation into the soft tissues The meningocele rather than the bifid spinous process is the important element The condition is a congenital malformation consisting of a defect in the closure of the vertebral arch usually with an associated defect of the meninges and their tissues There is in most instances no external meningeal sac although there is frequently a localized growth of hair Frequently there is a lipoma directly beneath the skin or in the deeper structures In practically every instance, the central portion of the cauda equina is fused into a single irregular bundle which may be the site of various pathological processes involving the nerve roots with resultant disturbance of function The commonly associated lesions comprise lipomas dermoid cysts, and bands of dense connective tissue There may be paralysis from the time of birth or the symptoms may not develop until later in life A traumatism may precipitate the symptoms The usual manifestations are lack of control of the bladder, toe drop, and saddle anesthesia Anterior sacral meningocele presents no abnormal manifestations on the external surfaces of the body With a large hernia there is pressure on the rectum bladder and other pelvic organs The common complaints comprise constipation frequency urgency and dysuria Dysmenorrhea and dyspareunia may be present Headache is a prominent symptom particularly in women The headache may develop during or subsequent to evacuation of the bowel It has been postulated that the headache is the result of pressure on the pelvic mass with temporary variation of the intracranial pressure, the changes in intracranial pressure being due to transmission of dynamic alterations through the spinal canal Physical examination is usually negative although in some instances the hernia may be palpable as a soft, fluctuant mass in the pelvis in the region of the upper portion of the posterior rectal wall Surgical amputation of the hernial sac is a major procedure and should be essayed only if the patient is incapacitated or presents other severe symptoms Aspiration of the hernial sac through the wall of the rectum has been performed as the result of a mistaken diagnosis of perirectal or pelvic abscess This usually results fatally due to meningitis

Roentgen Manifestations The characteristic roentgen manifestation comprises a hiatus of varying size and shape involving the anterior aspect of the sacrum The agenesis of the anterior segment of the sacrum may be unilateral or bilateral and usually affects the inferior and middle portions of the bone The coccyx is absent Remnants of the pedicles and laminae may remain The defect may be more marked on one side than the other Absence of the normal bony structures in the sacrococcygeal region is pathognomonic and permits of a definite diagnosis in most instances Other developmental anomalies such as lumbarization of the



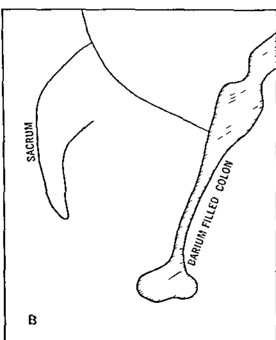




FIG 105 Anterior Sacral Meningocele A Anteroposterior view There is an extensive area of bone destruction involving the lower third of the sacrum The margins of the area of rarefaction in the sacrum are irregular in outline and poorly defined There is complete absence of the coccyx B Sketch of Figure C C Barium Enema lateral projection The lower portion of the sacrum and coccyx is absent The ampulla of the rectum and the sigmoid are displaced anteriorly and show marked narrowing (arrow)

first sacral segment and spina bifida may be present The hernia may be visualized as a rounded area of soft tissue density in the mid pelvis The mass lies at a higher level than the bladder Lateral views are of value to outline the soft tissue density in the pelvis Opaque enema studies show narrowing and displacement anteriorly of the ampulla of the rectum and the sigmoid The descending colon may be distended and fail to evacuate previously injected opaque material, the postevacuation roentgenogram showing practically total retention The hernia may lie to one side of the mid line with resultant displacement of the rectosigmoid to the opposite side Opacification of the bladder and the ureters reveals pressure deformities and displacements of these structures Myelographic studies afford definite proof of the lesion Injection of opaque material into the lumbar subarachnoid space results in migration of the opaque material into the sac of the hernia, demonstrating its size, shape and location It is essential that the patient be examined in both the erect and recumbent positions with anteroposterior, posteroanterior, lateral, and oblique projections

Differential Diagnosis In differential diagnosis it is necessary to consider tumors of the sacral canal, neoplasms of the sacrum, and pelvic masses of various types Tumors arising within the sacral canal, of which the ependymal cell glioma is the most common, cause smooth erosion of bone The bony defect may be solitary or multilocular Decalcification of the neural arches due to bone erosion is usually absent in occult meningocele Neoplasms arising within the sacrum are most commonly chordomas Ewing's tumor and osteogenic sarcoma present the usual characteristics of malignant bone lesions A benign giant cell tumor is characterized by trabeculation and deformity of the bone outline, although in sacral meningocele trabeculae may be preserved throughout the bone defect and closely simulate the appearance of giant cell tumor Pelvic masses and tumors originating in structures adjacent to the sacrum anteriorly or posteriorly particularly teratomas do not produce similar changes and are usually diagnosed without difficulty The presence of the anterior defect differentiates the anomaly from the more common posterior sacral meningocele which is due to failure of fusion of the posterior sacral arches and is characterized by the presence of a hernia at the posterior surface of the body

Spina Bifida Associated with Dislocation of the Hip and Other Anomalies

The association of dislocation of the hip with spina bifida is a well-established entity Spina bifida comprises a congenital, developmental defect of the neural canal characterized by imperfect closure of the spinal lamina Associated abnormalities of the skin subcutaneous tissues meninges and spinal cord occur frequently Spina bifida is most common in the lumbar and sacral regions but may occur in any portion of the spine It may be accompanied by other anomalies the most important of which are anencephaly hydrocephaly talipes equinus, harelip and cleft palate Myelocele meningomyelocele and spina bifida occulta comprise variations in the severity of involvement Patients with very marked spina bifida seldom survive for long periods unless the lesion is corrected surgically As the abnormality most commonly occurs in the lumbosacral

region, the lower limbs are usually affected Frequently one group of muscles is of good power while the antagonistic muscles show complete paralysis Sphincter disturbances are common There are two types of congenital dislocation of the hip The endogenous form is the result of developmental arrest of the acetabular roof The second is termed the dynamic type and is caused by abnormal forces which displace the femoral head from the acetabulum There may be a paralytic factor with relaxation of the ligaments about the hip and muscle imbalance

Chapter

3

Traumatic Lesions

In Collaboration with IRVING A. SHAUFFER, M.D.

FRACTURES

Introduction. Fractures of bone are caused by a trauma of short duration. If the bone is of normal structure only forces many times greater than the physiologic or forces exerted in an unphysiologic direction can produce a fracture. This type of fracture is termed traumatic. If the bony structure has been changed by disease such as senile osteoporosis, rickets, osteomalacia, osteomyelitis, hyperparathyroidism, tumors, and similar conditions, even physiologic stresses may lead to a fracture. In these instances it is termed a pathologic or spontaneous fracture. A fracture is described as complete or incomplete according to whether it leads to complete or partial discontinuity of the bone. In the case of a complete fracture the fragments may be either approximated or displaced. Severe injury to a bone leads to a comminuted fracture, characterized by splintering at the site of impact with smaller or larger fragments of bone between the two main fragments. When the site of fracture communicates with the outside through a wound of the overlying parts the fracture is termed compound or open. A fracture which is protected by the skin is termed simple or closed.

A fracture is a break in the continuity of a bone and as a rule is shown on the roentgenogram by a shadow of increased radiance extending partially or completely across the bone. Since the fracture line or lines permit the rays to pass through more easily than normal bone, the fracture line is seen on the roentgen film as a darker shadow. In the case of impaction or overlapping of the fragments, the fracture is manifested as a shadow of increased density. Separation or displacement of the fragments is demonstrable on roentgen examination. The various types of fractures such as complete, incomplete, torus or buckling, linear, oblique, stellate, spiral, etc., may be diagnosed with accuracy on the roentgen film. The extent of the fracture and complications such as extension into nearby joints or dislocation may be determined. Associated injury to the soft tissues is also demonstrable by the roentgen study.

Greenstick fractures may show no definite break in the continuity of the bone, being diagnosed merely by a slight increase in the density of the bone. This type of fracture is commonly seen in young children and is easily overlooked unless care is exercised in diagnosis. Sharp films with complete elimination of motion are essential. In some instances even simple linear fractures of the flat bones are difficult to demonstrate. Films

in the conventional sagittal and lateral projections may not reveal the fracture, oblique or tangential views being essential This is particularly true in the case of the carpal scaphoid, the tarsal bones and the skull The wrist should always be examined in three different planes Even with the most careful technique, the fracture may not be demonstrated at the examination immediately after the traumatism If the symptoms persist, it is necessary to re examine the patient after the lapse of a few days or weeks With the passage of time, absorption takes place along the line of fracture, slight displacement may result, or callus may be laid down This is particularly true in the case of the transverse processes of the lumbar vertebræ and the ribs a subsequent re examination often revealing one or more fractures which were not demonstrable at the first examination Distortion, compression, or irregularity of the bone trabeculæ is important in the diagnosis of fracture particularly in the flat bones and the cancellous portions of the long bones Fracture lines may be obscured by overlying structures, especially thin linear fractures without separation Fractures with overlapping of the fragments or impaction are manifested on the roentgenogram as areas of increased density

The position of the fragments is demonstrable on roentgen study and should be described anatomically in each instance with reference to either the proximal or distal fragments or both Angulation is designated in degrees and the direction of the angulation noted Shortening or overlapping may be estimated but allowance must be made for the magnification or distortion inherent in the roentgenogram If joints or other important structures are involved this must be described in detail as the prognosis is thereby materially altered

The treatment of fractures may be aided by the roentgenologist as he can determine whether the fragments are in satisfactory apposition and alignment or whether manipulation is necessary The opinion of the roentgenologist is valued in direct proportion to his knowledge and understanding of the situation Therefore it behooves every worker with the roentgen ray to be well grounded in not only the diagnosis but the reduction healing and care of fractures Films taken in several planes are essential Fluoroscopy may be of importance but should be performed only by an experienced roentgenologist as serious injury to the patient and surgeon may result from careless or inexpert techniques Perfect mechanical alignment is not always necessary although it is desirable and should be obtained if possible In the weight bearing bones the lines along which the body weight is transmitted may be determined on the roentgen film and may be more important than anatomical lines Overriding of the fragments for a distance of two centimeters or less is also satisfactory in the case of the long bones If one third to one half of the surfaces of the fractures are in apposition a good result may be anticipated The trauma incidental to repeated reductions must be given due consideration in determining whether further manipulation is essential Nature forms new bone on the side of the greatest defect so that even with apparently poor alignment of the fragments a good result is often obtained After the passage of time a new medullary cavity is re established in the case of the long bones In children after an interval of years it may be impossible to determine that a fracture had ever occurred In the case of fractures of the shafts of both bones of the lower legs the fact that the fragments of the tibia and the fibula are in contact is of no great practical significance With fractures of both bones of the forearms, synostosis of

the radius and ulna may be very serious as it prevents pronation and supination Maintenance of the position of fractures is of the utmost importance and fractures which are in good apposition must be maintained in such position and subsequent deformities due to muscle pull and weight bearing prevented Thus, a fracture of the spine may undergo increasing compression and angulation if the body is not placed in extension or a fracture of the ribs may cause injury to the lung or pleura unless held in place and all motion of the fragments prevented

Splints and heavy casts may obscure fracture lines or make it impossible for the roentgenologist to express an opinion as to position, callus formation, and healing Exposures at different angles or with varying degrees of penetration of the rays may overcome this difficulty If not the examiner must inform the clinician and a decision reached as to the advisability of removing the appliances Casts when wet obscure bone detail, less so when dry Care must be exercised to prevent tissue damage from excessive x-ray exposure and the number and frequency of re examinations is important The possibility of pathologic fractures must always be borne in mind Usually, fractures of the long bones are oblique or spiral, although transverse fractures may result The latter type must be viewed with suspicion as they are prone to occur in Paget s disease and malignancy of the bones Any unusual thickening, bowing or rarefaction of the bones should raise the possibility of a pathologic fracture As many pathologic fractures unite slowly if at all, the prognosis must be guarded in the presence of underlying disease of the bone

Time of X-ray Examination While it is usually customary to perform roentgen examination as soon as possible after the injury, in some instances this is inadvisable and it may be preferable to delay the x ray studies In skull injuries, the prevention of hemorrhage and shock may be as important as the demonstration of the fracture With injuries to the spine similar precautions are advisable to prevent further trauma which may result in paralysis or death Fractures of the ribs or sternum may cause serious trauma to the lung or pleura if the patient is moved Recent injuries referred to the roentgenologist without the application of a dressing must be handled with extreme care to prevent further damage or shock by motion of the fragments

Age of Fracture While there seldom arises a question as to the duration of a fracture, the determination of this fact may in certain instances become a matter of the utmost importance There are definite roentgen signs which are significant in this regard A recent fracture shows sharp, distinct clear cut fracture lines swelling and thickening of the soft tissues in the region of the injury, and absence of atrophy of the bones After the fracture is about a week old, the fracture line is less sharp the soft tissue swelling has disappeared, and there is slight or moderate atrophy In very young children callus may begin to form within five to ten days Within two to three weeks, there is definite filling in of the fracture line, bony callus is usually visible and there is atrophy of the bones and soft tissues After the lapse of four to six weeks the fracture line shows progressive obliteration the atrophy lessens, and the callus increases in size and density Old, healed fractures are characterized by bony thickening at the site of fracture and little or no bone atrophy In cases in which the fracture has failed to unite or there is delayed union, the sequence of the healing process is delayed or altered Fractures of the flat bones require weeks or months longer for union than is the case with the long bones

Roentgen Aspects of Healing of Fractures. In all fractures, frequent roentgen examinations are indicated to study the progress of healing. Healing is manifested by increasing indistinctness of the fracture line and the formation of bony callus. In the young new bone formation may develop within five days to a week. Adults require at least two to three weeks before callus formation is noted. In some regions preliminary bone resorption occurs prior to healing and in these instances fractures which are poorly visualized if at all at the original examination immediately after the trauma become clearly visible after an interval of a few days or a week. Rib fractures frequently fall into this category. With certain fractures several months must elapse before new bone is laid down and union is effected. At times union does not take place and the fracture line persists indefinitely. The development of a bony callus is dependent on the presence of a wide marrow space. In certain flat bones in which this is lacking callus does not form. Fractures of the bones of the skull, particularly of the vault, are retarded in healing and show reduction in callus formation. In many instances the fragments do not undergo bony union, becoming united by dense, connective tissue. The fibrous union cannot be regarded as evidence of failure of functional healing. The periosteum is of great importance in the process of bone repair. Periosteal reaction is manifested roentgenographically as a thin dense line parallel to the long axis of the bone and separated from the latter by a narrow band of radiance. The radiant area decreases in width and eventually disappears, indicating that bone formation has ceased. With the lapse of time, the callus decreases in extent and the medulla usually resumes its continuity. In some cases the fracture line disappears completely or remains only as a narrow area of density. The extent of overriding and the apposition of the fragments are important in evaluation of the healing of a fracture. Involvement of a joint surface by the fracture is particularly important. Displacements of epiphyses are very significant and should be evaluated carefully in every instance. Fractures of the femoral neck are frequently treated by means of nails or pins inserted through the greater trochanter into the neck and head of the femur. Fluoroscopic control is frequently utilized in this procedure. Plating, wiring, intramedullary nails, and prostheses of various types are coming into widespread use. The roentgenologist must be familiar with the different methods and procedures. Stereoscopic studies must be used in the case of the shoulder, hip, and other regions in which two projections at right angles to each other cannot be obtained. Comparative studies of the unaffected side in positions corresponding to those utilized in the examination of the injured part are of great value, particularly in children. Laminography and similar diagnostic aids should be used whenever indicated.

Histologic Aspects of Healing of Fractures. The anatomical findings in fractures are dependent on hemorrhage and the changes which develop as healing progresses. The rupture of blood vessels in the bone marrow, the periosteum, and the adjacent muscles causes the development of a hematoma in the region of the fracture. The bleeding extends into the bone marrow and the surrounding soft tissues. The blood of the hematoma coagulates within six to eight hours after the injury. Later, the soft tissues in the adjacent region show active hyperemia and edema. The blood clot undergoes organization and is gradually transformed into a callus. The callus increases in size during the period of four to six weeks after the injury and then gradually diminishes and disappears completely

with the passage of time In those cases in which the fracture is in good position and healing progresses normally, the site of the fracture may not be demonstrable after a lapse of three to four months The fragments in many instances are mobile The mobility diminishes with the development of the callus

In most instances the healing of a fracture involves the formation of a cartilaginous callus Membrane bones heal without cartilage formation Once the formation of bony callus has begun, the progress of its formation can be followed by successive roentgen examinations because the mature bone has a greater mineral content than the immature bone During the formation of callus, an excessive amount of bone is produced This serves as a protective measure The volume and amount of callus are greatest at the concavity in cases in which the bone fragments are not in perfect alignment This serves to provide protection against further angulation and displacement of the fragments and is probably the result of bending forces acting upon the callus which is in the process of being formed After solid union of fragments which are in good apposition and alignment the surplus bone formed at the site of the fracture as a thick spindle loses its functional importance It is gradually reduced and finally disappears completely by resorption This process of resorption of bone which is not under mechanical stress is in conformity with the development which takes place in the normal skeleton After a fracture in good position has undergone complete healing, the site of the fracture often cannot be detected by gross anatomic or roentgen studies In many cases the alignment of the fragments is not perfect although sufficiently good for clinical and functional result In these instances the bone and the callus are remolded according to the functional stresses Some portions of the callus are removed and other portions are involved in the process of internal reconstruction, being incorporated into the reshaped bone The original fragments are molded by resorption and apposition These changes may extend into areas at a considerable distance from the site of the fracture and also far beyond the enveloping callus The functional reconstruction of the callus is a slow process and even under the most favorable conditions may continue for months and years

The Healing of Pathologic Fractures A pathologic fracture is a fracture occurring in a diseased bone The fate of such a fracture is dependent upon the underlying pathologic process The pathologic changes may be local and restricted to one bone or segment of bone or systemic in origin and involve multiple bones or the entire skeleton Osteoporosis may be generalized as in old age and certain metabolic diseases or confined to a single bone or a single group of bones as in disuse Senile osteoporosis is the result of lessening of bone formation and is a manifestation of the decreased cellular activity of old age Callus forming after fracture of a bone which has been rarefied by hyperparathyroidism is subject to the same pathological influences as the entire skeleton The callus forms in practically normal amounts and at normal speed However, the bone thus formed soon falls prey to resorption The presence of primary or metastatic tumors may weaken a bone to such an extent that it fractures under physiologic stress Callus formation is dependent on the character of the underlying neoplasm and other factors Tumors which expand and progress slowly show satisfactory healing as a rule In the presence of a rapidly growing tumor, callus formation may be prevented or delayed Rickets is characterized by lack of calcification of cartilage and of newly

formed bone The bone matrix formation is not impaired The changes in the callus after fracture of a rachitic bone are in every respect similar to those which occur in the entire skeleton Fibrous and cartilaginous callus develop as normally The ossification of the fibrous callus proceeds normally in so far as the formation of bone matrix is concerned The osteoid tissue does not calcify The formation of endochondral bone which should replace the cartilaginous callus does not occur because the hyaline cartilage fails to calcify and is not removed by the proliferating osteogenic tissue

The Healing of Fractures Associated With Osteomyelitis In a compound fracture, healing begins simultaneously with the healing of the soft tissue wounds and proceeds in the same manner as in simple fractures If the compound fracture becomes infected the process of healing is disturbed This is due principally to damage to the tissues at the site of the fracture and is manifested by the development of a purulent periostitis and osteomyelitis The resorption of the necrotic bone and the apposition of new bone are retarded or inhibited The formation of new bone proceeds only at the periphery of the infected area and may be excessive In the presence of osteomyelitis, this leads to the formation of an involucrum Resorption of bone begins at the boundaries between the necrotic and the undamaged bone with formation of a sequestrum Small, isolated fragments of bone undergo necrosis and either extrude or must be removed surgically The formation of callus may begin after the acute infection has subsided or when the process has become chronic In cases of prolonged infection or extensive loss of bone bony union does not occur and pseudarthrosis develops Pseudarthroses may also develop because of excessive mobility of fragments interposition of soft tissues between the fragments and a decrease in the regenerative capacity of the patient such as occurs in old age, syphilis and other diseases The simple type of pseudarthrosis is characterized by fibrous union of the fragments In cases of long standing the surfaces of the fragments become smooth plates of compact bone Because of the influence of shearing forces at the site of the fracture the opposing surfaces of the fragments of bone may become covered by a layer of fibrocartilage or hyaline cartilage which is differentiated from the connective tissue between the fragments The presence of cartilage in a pseudarthrosis must be regarded as a step in the functional adaptation to a new pathologic joint

Fracture of the Clavicle

Fractures of the clavicle may involve any portion of the bone but are most frequent in the middle and outer thirds The lateral fragment is usually displaced downward and inward Greenstick fractures without displacement and fractures with bowing usually upward at the site of the fracture are common The complete fracture with displacement is very difficult to reduce satisfactorily and in girls or young women the over riding of the fragments may be a cause of concern because of the deformity which may ensue Union is usually satisfactory and normal function is re established in most cases

Fracture of the Scapula

Fractures of the scapula are rare and are often difficult to demonstrate Any portion of the bone may be involved Fractures of the neck the

acromion process and the body occur most commonly. Separation and displacement of the fragments are demonstrated by oblique projections or stereoscopic studies. If the arm can be elevated over the head, lateral projections are obtained by rotating the patient slightly toward the affected side.



FIG. 106. Fracture of Clavicle. The fracture occurred three weeks previously. There is extensive callus formation about the margins of the fracture.



FIG. 107. Fracture of the Scapula. There is a fracture involving the glenoid and the adjacent portion of the body of the scapula. The fragments are separated. The patient fell from a height landing on the shoulder.

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or more ribs, re-examination three to six weeks later disclosed an additional fracture of the ribs in 32 cases, 2 more in 9 cases, and 3 more in 1 case In 6 cases with a single rib fracture, the fracture could not be identified at a subsequent re-examination In 9 cases with multiple fractures, re-examination demonstrated one less fractured rib than at the original study Since re-examination after an interval of a few weeks failed to show a fractured rib although the lesion could be identified on the original examination, it appears that approximately 15 per cent or more of rib fractures heal without periosteal callus and become unidentifiable at a subsequent study This accounts for the fact that many cases with fracture of the ribs are undiagnosed on roentgen examination made immediately after injury, while in other cases, rib fractures heal so completely that the fracture cannot be visualized a few weeks or months after the trauma

There are several reasons for the difficulty in the demonstration of fractures of the ribs immediately after the injury The ribs are relatively small structures compared with the mass of tissues which the rays must traverse This is particularly true in the case of the lower ribs Only about one third of the rib can be brought into close approximation to the film on any single exposure The remaining portions of the ribs are in different planes may lie in the same axis as the central ray and may be at a considerable distance from the film Because of the different densities of the structures of the chest the upper ribs cast a much darker shadow than the lower ribs The inferior ribs show relatively slight contrast due to the muscular tissues with which they are surrounded Unless the injury is severe the fragments are not separated and present smooth margins After the lapse of three to six weeks, the fragments may separate the edges become ragged, or callus forms, giving conclusive evidence of the presence of fracture In the presence of clinical evidence of fracture of the rib even though the roentgenogram is negative, the patient should be re-examined three to six weeks after the injury This will greatly reduce the number of errors in the roentgen diagnosis of rib fractures An important aid is for the clinician to designate as exactly as possible the ribs and the portions of the ribs most probably involved It is necessary to utilize multiple projections and various exposure techniques For the first to ninth ribs the exposure is similar to that for the chest It is helpful to use the Bucky diaphragm For the lower three ribs, kidney or the lumbar spine techniques are best

Bloom has suggested a method of increasing the accuracy of the diagnosis of rib fractures It is termed the "continuous respiration technique" Liberson suggests a single film, two exposure technique The first exposure is the usual anteroposterior view The second is made as follows On the table top three lines are drawn one median the others three inches to each side of the midline The patient lies supine and is placed so that the side of the chest to be examined is centered on the midline of the table A right angle is placed on the lateral ruled line of the table The involved side of the chest barely touches the vertical bar of the right angle at about the ninth rib The patient is strapped with a compression bladder and breathes naturally, preferably with the diaphragm The exposure is a slow prolonged one accomplished by using only 10 milliamperes The peak kilovoltage is proportionate to the thickness of the patient usually the same as for the study of the kidneys About six to eight seconds exposure time suffices to blot out the vascular

Fracture of the Sternum

Fracture of the sternum was formerly a very rare condition In recent years, it is being seen with increasing frequency and is usually the result of the driver of an automobile being thrown against the steering wheel in a collision The fracture usually involves the body and is of the oblique or transverse type Separation and displacement as a rule are slight Comminution is rare The lesion is difficult to demonstrate because the bone is obscured by the overlying mediastinal structures and the shadow of the spine in the sagittal projections and also because it is painful for the patient to lie in the prone position Oblique and lateral views are essential to determine the position of the fragments Laminography is of great value and should be utilized in all doubtful or suspicious cases



FIG 108 Fracture of the Sternum Laminography Lateral View There is an oblique fracture of the upper third of the body of the sternum with overlapping the distal fragment being displaced backward and upward Routine roentgenograms did not demonstrate the fracture clearly The laminographic studies established the diagnosis definitely and showed the exact relation of the fragment

Fracture of the Ribs

The roentgen diagnosis of fracture of the ribs following a chest injury is entirely satisfactory when a fracture is demonstrated Every radiologist has had the trying experience of performing careful roentgen study of ribs with no evidence of fracture on the first examination yet a few weeks later to have re examinations reveal one or more fractures which could not be identified in the original roentgenograms even though their exact location was known Re examination at a subsequent date is essential before a negative diagnosis of rib fracture is established The inadequacy of a single roentgen examination in the demonstration of rib fractures is illustrated in a report by Liberson In 100 cases of recent chest injury suspected clinically to have rib fractures in which roentgen examination was negative re examination three weeks later disclosed fractures of the ribs in 22 cases In a second series of 100 cases with fracture of one

or more ribs re-examination three to six weeks later disclosed an additional fracture of the ribs in 32 cases 2 more in 9 cases, and 3 more in 1 case In 6 cases with a single rib fracture, the fracture could not be identified at a subsequent re-examination In 9 cases with multiple fractures, re-examination demonstrated one less fractured rib than at the original study Since re-examination after an interval of a few weeks failed to show a fractured rib although the lesion could be identified on the original examination, it appears that approximately 15 per cent or more of rib fractures heal without periosteal callus and become unidentifiable at a subsequent study This accounts for the fact that many cases with fracture of the ribs are undiagnosed on roentgen examination made immediately after injury, while in other cases, rib fractures heal so completely that the fracture cannot be visualized a few weeks or months after the trauma

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and bronchial structures of the lungs as the patient breathes during the exposure. A 14 × 17 inch film is placed in the Bucky tray and one half of the film is covered by a 7 × 17 inch strip of lead. The uncovered part of the roentgenogram is centered *in the middle of the tray*. The exposure is made during inspiration and expiration. The film is then shifted so that the unexposed part is centered and the lead is placed over the exposed half of the film. If the Bucky tray does not accommodate the shift of the cassette, the cassette is turned 180 degrees and the patient also turned so that the head and feet exchange positions. The patient next lies on the abdomen so that the side of the chest to be studied is centered on the unexposed half of the film. During this exposure, the patient suspends respiration at the end of inspiration and a short exposure time is used. Each half of the film must be placed accurately and the patient carefully centered. All twelve ribs are seen in the majority of cases. This technique has many advantages. There is a great increase in the percentage of correct diagnoses. Since one exposure is made during respiration the shadows of pulmonary vessels and bronchi are obliterated by the movements of these structures during the long exposure. The study affords a clear outline of the posterior and the axillary parts of the ribs. The suspended respiration posteroanterior view affords clear definition of the anterior portion of the ribs.

The common complications of fractures of the ribs are pleural effusion, pneumothorax, hemothorax, atelectasis and subcutaneous emphysema. Roentgen study of the chest in addition to the examination of the ribs is essential to establish the diagnosis of complications which may occur in association with the fracture.

Cough Fractures of the Ribs. Fractures of the ribs after coughing are not infrequent. The disease is particularly common in patients with pulmonary tuberculosis. There is usually a history of pain following violent coughing or sneezing. Cases have been recorded late in pregnancy, all of which occurred during fits of coughing. The mechanism of the fractures is uncertain. Abnormal fragility of the bones is not considered a contributing factor. The lesion is most probably associated with asymmetry of posture with resultant inequality of the ribs. Prolonged bed rest with incident decalcification may be a contributory factor. During pregnancy the fractures are most probably due to mechanical changes associated with protuberance of the abdomen and compensatory lordosis of the body. The fractures are similar to march fractures, spinous process fractures in earth shovelers and similar fatigue fractures. Primary osteoporosis due to avitaminosis and pulmonary disease of long standing are predisposing factors. The patient may be unaware of the fracture and it may heal with practically no symptoms.

Fractures of the First Rib. Isolated fractures of the first rib are rare. In order to understand the mechanism of these fractures, a knowledge of the anatomy of the first rib is essential. The first rib is situated deeply about in the center of the neck. It is protected by the clavicle anteriorly and the scapula and the muscles of the shoulder girdle posteriorly. The superior surface has two shallow depressions separated by the *tuberculum scaleni* to which is attached the scalenus anticus muscle. The subclavian vein passes over the groove in front of the tuberculum and the subclavian artery passes behind it. There are three muscles attached to the first rib: the scalenus medius, the scalenus anticus, and the serratus magnus. Because of its anatomical location it is unlikely that direct trauma can affect

the first rib alone, although isolated fractures of this rib may occur With indirect trauma, the force is transmitted through the manubrium or the clavicle A direct trauma to the back may be the etiologic factor Transmission of force through the clavicle is the most common cause, the shoulder suddenly being moved downward and backward and the clavicle striking the first rib In many cases of isolated fracture of the first rib, no history of trauma to the chest can be elicited It is believed that a sudden stress with contraction of the scalenus anticus muscle is the cause Not infrequently, there is an appearance which simulates a fracture of the first rib on the roentgenogram Many of these are not actually fractures and are due to an anomalous development of the first rib Only 263 cases of isolated fracture of the first rib have been recorded in the literature and of these but 17 were bilateral The majority are found during radiography of young healthy adults particularly in the armed services and are comparable to the stress or fatigue fractures which occur in the metatarsals tibia fibula and other bones In most cases the patients are unable to remember any injury or symptoms attributable to a fracture although in a few there is a history of a pain in the neck and shoulder on lifting or straining The fracture is usually asymptomatic at the time of roentgen examination

Roentgen examination may reveal a linear crack in the rib or a pseudarthrosis with widened irregular, sclerotic opposing surfaces In many instances, the fracture is found to be firmly healed with varying degrees of thickening at the fracture site Serial roentgen studies show that nonunion or formation of a false joint often follows this fracture, although bony union does occur in many instances Many authors believe that this lesion is not a fracture and consider it a developmental anomaly particularly because of the absence of a history of injury and the rarity of ununited fractures of other ribs The body of the first rib may arise from two centers of ossification and the junction may persist as a synchondrosis However this is more posteriorly than the usual site of fracture The line of division between the two bony segments lies in a sagittal plane and not transversely as with fracture The sex incidence favors an acquired lesion An earlier radiograph may be available and show a previously normal rib Fractures of other ribs particularly the seventh and eighth are frequently caused by sudden muscular effort and severe coughing In the lower ribs the fractures usually develop at a point where two muscle masses pull in different directions This type of fracture in some instances has led to death from rupture of the adjacent vessels *Fracture of the first* rib may be associated with fractures of the clavicle, scapula, spine or other ribs and is often complicated by injury to the subclavian vessels, the brachial plexus, or the pleura

Fracture of the Humerus

A Fractures of the greater tuberosity are *important* because the separated fragment may be confused with a calcified subacromial bursa The rough irregular fracture line establishes the diagnosis

B Fracture of the surgical neck is the most common of the fractures of the upper end of the humerus The tuberosity may be fractured simultaneously When lateral views *cannot* be obtained, stereoscopic studies should be carried out in order to permit of accurate evaluation of the position of the fragments

C Fracture of the anatomical neck is rare but is of the utmost importance as marked rotation and displacement of the head may occur These fractures are in many instances intracapsular and delayed union or non union is apt to result

D The shaft of the humerus may be broken at any level Comminuted, oblique or spiral fractures are common Displacement may consist of bowing or overlapping of the fragments Lateral bowing must be corrected as healing in this position results in a bad carrying angle with severe disability Slight medial bowing does not interfere with function



Fig 109

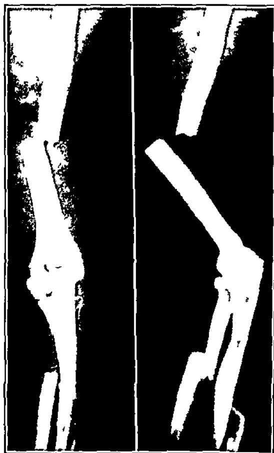


Fig 110

Fig 109 Comminuted Fracture of the Humerus There is a markedly comminuted fracture of the humerus in the region of the upper and middle thirds of the shaft with separation and displacement of the fragments

Fig 110 Fractures of the Shaft of the Humerus and Both Bones of the Forearm with Marked Displacement of the Fragments

E Supracondylar fractures of the humerus are among the commonest injuries in the region of the elbow joint Lateral or posterior displacement of the distal fragment is frequent Involvement of the articular surface of the humerus is of importance in prognosis

F Fractures of the condyles may be widely separated and require operative replacement Studies in various projections may be required to demonstrate the fracture Films of the unaffected side are of great aid in demonstrating the type of fracture and the degree of displacement

Fracture of the Radius

A *Head* Fractures of the head of the radius may be extremely difficult to demonstrate as the patient is unable to fully extend the elbow With the elbow in partial flexion the roentgen study requires several different projections, one with the upper forearm on the film and another with the lower end of the upper arm in apposition to the roentgen film Early recognition of fracture of the radial head is essential as failure to immobilize the elbow may result in displacement of the fragments and marked impairment of pronation and supination of the forearm Excessive callus formation and non union are indications for operative intervention



Fig 111 Comminuted Fracture of the Lower Third of the Bones of the Forearm as Visualized Through a Plaster Cast

B *Neck* Fractures of the neck of the radius require no special techniques and are easily demonstrable

C *Shaft* Any portion of the shaft may be fractured Oblique views are helpful to show whether the fragments are in contact with the ulna In fractures of both bones of the forearm, synostosis of the radius and ulna must be prevented because of interference with pronation and supination

D *Lower End* The commonest injury of the lower end of the radius is the comminuted impacted fracture termed Colles' fracture Shortening and backward displacement of the fragments cause serious impairment of function and must be corrected unless reduction is contraindicated because of the age or condition of the patient Lines may be drawn on the

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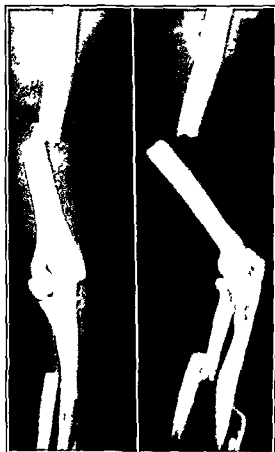


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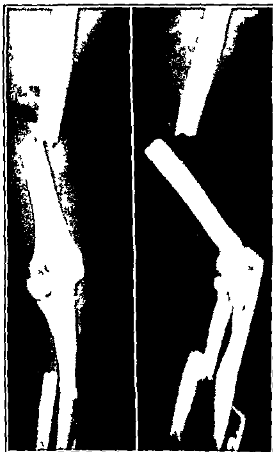


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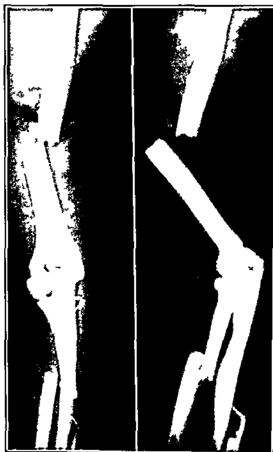


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E Supracondylar fractures of the humerus are among the commonest injuries in the region of the elbow joint Lateral or posterior displacement of the distal fragment is frequent Involvement of the articular surface of the humerus is of importance in prognosis

F Fractures of the condyles may be widely separated and require operative replacement Studies in various projections may be required to demonstrate the fracture Films of the unaffected side are of great aid in demonstrating the type of fracture and the degree of displacement

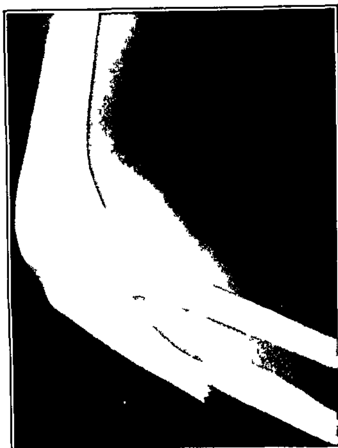


Fig 113 Fracture of the Upper Third of the Shaft of the Ulna

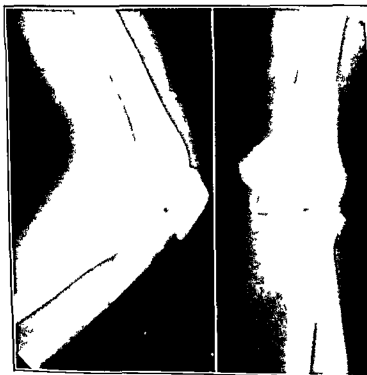


Fig 114 Fracture of the Olecranon Process

film indicating the long axis of the shaft and the fragments and to show the tilting of the articular surface to assist in the determination of the position of the fragments

Greenstick and subperiosteal fractures are frequent in children and are easily overlooked unless the roentgenograms are of excellent quality and are studied carefully In children with separation of the lower radial epiphysis, there often occurs an oblique fracture of the posterior aspect of the lower end of the diaphysis of the radius the fragment being displaced backward and upward with the epiphysis Fracture of the styloid process of the radius may occur Barton s fracture is a transverse fracture at the previous site of the epiphysis of the lower end of the radius

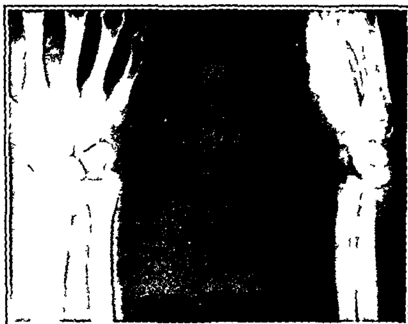


Fig 112 Colle s Fracture There is a comminuted impacted fracture of the lower end of the radius The articular surface of the radius is tilted backward

Fracture of the Ulna

A *Upper End* Fractures of the olecranon process are relatively common and separation of the fragments is the rule This is the only fracture in the region of the elbow which is immobilized with the arm in full extension Unless properly reduced the fragments do not unite Hence early diagnosis and prompt therapy are important Open reduction is required in the presence of wide separation of the fragments

Fractures of the coronoid process involve the articular surface of the ulna Posterior dislocation of the ulna usually occurs

B *Shaft* Any portion of the shaft may be fractured The upper one third is most commonly involved In this instance there is usually an associated anterior dislocation of the radial head Subperiosteal or green stick fractures occur in children In adults there is usually over riding or displacement Synostosis between the radius and ulna must be avoided to prevent loss of pronation and supination of the forearm

C *Lower End* Fractures of the styloid process may occur usually in association with fracture of the lower end of the radius (Colle s fracture)

Fractures of the Carpus

There has been no generally accepted series of positions utilized to supplement the standard posteroanterior and mediolateral projections in the study of the carpus In order to determine the minimum number of positions necessary to demonstrate all of the carpal bones and joints, the wrist was studied by Roderick He concluded that at least five projections are required to permit of complete study of the carpus (1) Posteroanterior The wrist is placed in pronation with ulnar deviation The central ray is directed perpendicularly to the plane of the wrist and one half inch distal to the mid point of the interstyloid line This results in an excellent view of the carpal bones and their interspaces The navicular and the greater multangular are shown with clarity The lunate the triangular the capitate the hamate and the carpometacarpal interspaces are well defined A slightly better view of the hamate is obtained without ulnar deviation (2) Oblique posteroanterior view with the wrist in the lateral position and the medial side in contact with the film The wrist is rotated internally in the direction of pronation until the volar surface is at a 45 degree angle to the plane of the film and the wrist is deviated in the direction of the ulna This position is particularly valuable in the visualization of the proximal row of carpals The greater and lesser multangular and the proximal portion of the capitate are seen better than in the routine position Although some superimposition exists this view demonstrates the articulation between the distal row of carpals and the first four metacarpals (3) Oblique anteroposterior with the wrist in the lateral position and the ulna next to the film The wrist is rotated externally in the direction of supination until the dorsal surface forms a 45 degree angle with the plane of the film and is deviated toward the ulna This position demonstrates the proximal row of carpals with a minimum of superimposition and illustrates the relationships of the proximal row of carpals to those with which they articulate The navicular lunate triangular and pisiform are well outlined (4) The lateral, mediolateral view This view affords a lateral view of the navicular lunate triangular greater multangular capitate and hamate (5) The axial view The forearm is placed in pronation and in contact with the film The wrist is extended The degree of extension is dependent upon the degree of mobility of the part, a 90 degree extension being obtained if possible This view is important for a supplementary examination of the pisiform the process of the hamate the lesser multangular and the greater multangular bones

Fracture of the Navicular Of the small bones of the wrist, the navicular most frequently suffers a fracture A fracture of the navicular in practically all instances is in the form of a transverse intra articular fracture Fractures of the other carpal bones usually occur as extra articular chip fractures although there rarely may result intra articular compression fractures especially of the os magnum or the trapezium Fractures of the navicular appear to develop for two reasons The first and most important is the fact that the bone presents an area of narrowing termed the waist The second is the anatomical location of the bone in both rows of the carpal bones On movement of the wrist the navicular moves not only with the proximal row but also with the distal row of the bones of the carpus On radial flexion of the wrist the long axis of the navicular is placed approximately at right angles to the long axis of the forearm Fractures of the

Patella Cubiti

Patella cubiti is a condition involving the elbow joint and is characterized by the presence of a patella like bone proximal to the olecranon process and within the investments of the triceps tendon It is not definitely known whether the anomaly represents a simple congenital malformation or is due to trauma Habbe has published a report on a case in which the traumatic origin is well authenticated The condition is asymptomatic and is usually discovered during roentgen study for other reasons Avulsion of the patella cubiti presents a definite clinical picture There is a history of an extension force applied to the elbow followed immediately by pain and loss of the power of extension A moderately large

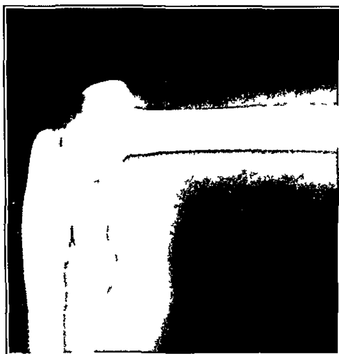


Fig 115 Patella Cubiti There is an area of bony density adjacent to the posterior aspect of the lower end of the humerus The appearance closely resembles that of a fracture of the olecranon process However there was no history of trauma and the calcific body is in the tendon

bony fragment is palpable posteriorly to the lower end of the humerus The fragment is movable but cannot be displaced distally as far as the elbow joint The aberrant osseous structure is classified as a sesamoid bone and appears to arise in the tendon of the triceps humeri rather than from a previously separated epiphyseal nucleus Roentgen studies show the avulsed sesamoid lying at a distance of 1 or 2 cm from the olecranon as contrasted to its usual position in immediate contact with the olecranon The margins of the bone are smooth The most important criterion for diagnosis is the fact that the combined length of the ulna and the sesamoid exceeds that of the normal ulna or if the condition is bilateral exceeds the expected length of the ulna is estimated from the radius Treatment consists of screwing or nailing the sesamoid to the ulna without destruction of the joint surface



Fig 116 Fracture of Navicular with Separation of the Fragments

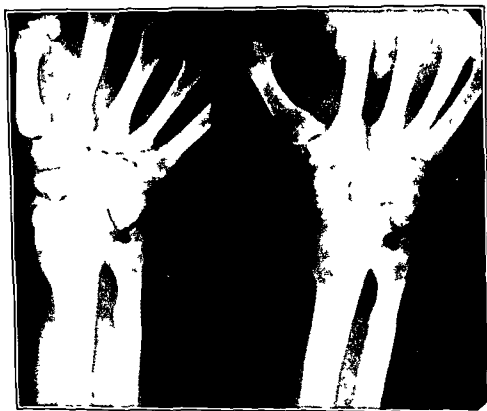


Fig 117 Ununited Fracture of the Navicular
Osteochondritis of the Fractured Navicular and the Lunate

navicular are of particular importance because of the great tendency to non union The blood supply is compromised by fractures of the proximal segment This tends to result in the development of avascular necrosis of the proximal fragment The roentgen manifestatious in this instance constitute an unchanged or, in some instances, increased density in the proximal fragment with marked osteoporosis of the distal fragment and the adjacent bones The portion of the bone which is covered with periosteum is very narrow The limited periosteal covering is an important factor in predisposing to fracture With separation of the fragments, the fragments may move separately, one moving with the proximal and the other with the distal row of the carpal bones It is difficult to retain the fragments in apposition and alignment Dislocation, partial or complete frequently results These and other factors enhance the tendency to non union

The lesion is most common in young males who do manual labor and is more frequent on the right The fractures are usually caused by indirect violence transmitted from the outstretched hand It is essential to use multiple views to demonstrate the fracture anteroposterior with the hand is marked ulnar deviation posteroanterior at a 45 degree angle oblique, lateral and oblique projection with the ulnar region elevated The fracture line may occasionally not be demonstrable at the first examination, only to become apparent on re examination two or more weeks later after absorption along the fracture line has occurred Slight separation of the fragments is common Displacement is rare except in the presence of associated anterior dislocation of the lunate Comminution is unusual Osteoarthritis frequently develops in cases of non union A single roentgen examination which reveals no demonstrable fracture must not be accepted as conclusive particularly if only routine anteroposterior and lateral views were utilized Early diagnosis and prompt therapy are essential if cure is to be obtained and operation avoided The common complications of fractures of the navicular are delayed union non union and aseptic necrosis Aseptic necrosis may be the result of crushing injuries or rough manipulation

Fractures of the Metacarpals and Phalanges

Fractures of the bones of the hand are relatively common They are most frequently oblique or spiral although transverse or comminuted fractures occur in crushing injuries Certain fractures of the bones of the hands are characteristic and the etiology may be determined by the character of the break in the bone This may be of importance in medico legal matters The distal end of the second and fifth metacarpal is often fractured during fighting The fracture shows impaction and bowing The articular surfaces are not infrequently involved A characteristic deformity results with loss of the prominence normally present at the knuckle if these fractures are not reduced The baseball fracture is a chip fracture of the dorsal aspect of the phalanx An oblique fragment of bone is broken usually at the joint Non union is common and a typical thickening and swelling eventuate

Bennett's Fracture Bennett s fracture is an intra articular fracture of the base of the metacarpal bone of the thumb The lower volar extremity together with a portion of the joint surface forms a fragment which is maintained in its correct position in relation to the greater multangular

FRACTURES OF THE LOWER EXTREMITIES

Fractures of the Femur

Head Fractures of the head are very rare and occur only as the result of severe traumas such as crushing injuries or falls from a height There is usually an associated fracture of the acetabulum and the neck of the femur and other injuries

Neck The neck of the femur is one of the most frequent sites of fracture It is of the utmost importance to determine whether the fracture is intracapsular or extracapsular fracture as the former type results in non union and absorption of the neck and head in many instances The attachment of the capsule is along the anterior aspect of the intertrochanteric line and is at a lower level along the posterior aspect of the bone than on the anterior surface Non union may result from interference with the blood supply, interposition of soft tissues or other causes The neck may be fractured at any point from the junction of the head and neck to the base The break may be oblique or transverse and is usually single, although comminution and accompanying separation of the lesser trochanter and a portion of the greater trochanter are frequent Stereoscopic views are important to show the displacement as lateral views may not be obtainable The usual displacement is with the shaft fragment upward and inward the angle between the neck and the shaft approaching a right angle The displacement may be determined in several ways The greater trochanter lies about 3 cm below and lateral to the superior border of the acetabulum In fractures of the neck, the upper portion of the greater trochanter lies nearer to the hip joint and may be at the same or even a higher level than the superior margin of the acetabulum A line drawn horizontally through the middle of the femoral head at the point of attachment of the ligamentum teres extends to the upper level of the greater trochanter and a line drawn vertically through the middle of the femoral shaft is normally about 4 to 5 cm lateral to the hip joint Shenton's line, formed by extending the arc of the inferior surface of the neck of the femur normally extends in a smooth curve along the superior border of the obturator foramen In fractures of the femoral neck, this line is displaced and irregular

Fractures of the neck of the femur are easily overlooked if the films are not technically perfect An impacted fracture cannot in some instances be visualized immediately after the trauma After the lapse of a few days or weeks absorption takes place along the line of *fracture* and the break in continuity becomes clearly visible Roentgen evidence of union is unreliable for the roentgenologist must wait until bony callus becomes visible This may not take place for many months Fibrous union may be sufficient to permit the patient to walk with the aid of an appliance long before callus is demonstrable on the roentgen film Fluoroscopic reduction and the insertion *of metallic nails* have become important adjuncts in the treatment of certain fractures of the neck of the femur The insertion of the nail is facilitated and accomplished with much greater accuracy with roentgenoscopic control Periodic roentgen studies are particularly important in studying the progress of healing The extent of the union can be demonstrated *the position* of the fragments observed and in cases of non union the absorption of the neck or head visualized

by the ligamentous attachments The shaft of the metacarpal bone is displaced dorsoradially due to the pull of the abductor pollicis longus muscle Unless properly reduced, the fracture results in a serious and permanent deformity In many instances, reduction and fixation can be accomplished only by operative procedures In the roentgen demonstration of this lesion a true lateral roentgenogram of the base of the first metacarpal and the joint between the first metacarpal and the greater multangular bone is necessary This view is obtained in the vertical plane by placing the palmar surface of the hand flat on the film and pronating the hand and wrist approximately 15 to 35 degrees If the degree of rotation is not sufficient, there is protrusion of the dorso radial tubercle of the greater multangular bone The roentgen tube is displaced distally and the central ray directed at an angle of 20 to 30 degrees from the vertical In this manner the ulnar and radial joint margins of the greater multangulum are superimposed upon each other In a true lateral projection, the first carpo metacarpal joint is projected over the base of the second metacarpal bone and the greater and lesser multangular bones become superimposed



Fig 118 Fractures of the Pelvis There is a fracture of the right pubis (black arrow) and a fracture of the ischium (white arrow) The fragments show slight separation

Fractures of the Pelvis

The bones of the pelvis are very frequently fractured The common causes are falls from a height crushing injuries and automobile accidents Comminution separation and displacement are the rule The iliac bones may show multiple fractures and the fragments are frequently widely displaced Involvement of the acetabulum is serious because of resultant limitation of hip movements The pubis and ischium are most frequently fractured in the region of the rami The pelvic ring is usually broken in two places and the fracture lines tend to be opposite each other The symphysis and acetabulum may be involved In the female narrowing or distortion of the obturator foramen and birth canal are of great significance and correction of deformities is of the utmost importance Separations of the symphysis pubis and sacro iliac joints may accompany these fractures and may be more significant than the injury to the bone itself

FRACTURES OF THE LOWER EXTREMITIES

Fractures of the Femur

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Fig 119 Fracture of the Neck of the Femur The shaft fragment is displaced upward and inward The angle between the head and the shaft of the femur is approximatley a right angle



Fig 120 Fracture of Neck of Femur with Metallic Nail *A* Anteroposterior view *B* Lateral projection *There is a complete fracture of the neck of the femur with slight impaction The nail is in good position*

Femoral Shaft Fracture of the shaft of the femur is usually oblique or spiral and may involve any portion of the shaft Transverse fractures occur less often and are usually the result of being run over by a team or automobile All transverse fractures must be viewed with suspicion as they are frequently pathologic Careful study is essential to rule out metastatic carcinoma, bone cyst, Paget's disease, or other underlying disease Reduction of femoral shaft fractures is difficult Metallic plates, pins, or bands are often utilized in the treatment of these fractures Supracondylar fractures frequently show marked displacement and separations Repeated examinations may be required to check the position and obtain proper reduction

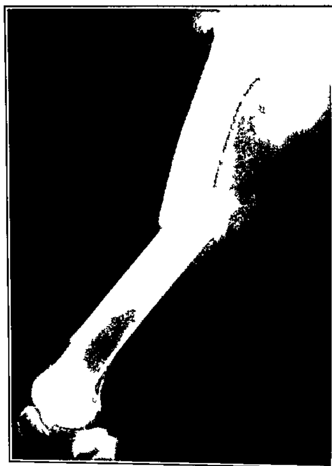


FIG 121 Fracture of the Mid Shaft of the Femur with Comminution Bowing and Overlapping of the Fragments

Fracture of the Patella

Fractures of the patella are usually transverse, although oblique or longitudinal fractures may occur They are best demonstrated in the lateral view as the patella may be practically completely obscured in the sagittal projection Oblique or other special positions may be required to demonstrate thin, linear or incomplete fractures A congenital anomaly known as partite patella may result in a division of the patella into two three or rarely more fragments This condition is frequently bilateral and comparison of both knees prevents error in diagnosis The margins of the fragments may be eburnated, dense and smooth in partite patella, while fractures usually present hazy, slightly irregular contours In recent

fracture there is fluid in the prepatellar bursa with marked swelling and thickening of the soft tissues about the knee The rapid development of osteoporosis aids in the diagnosis of fracture of the patella If the fragments are separated, union cannot take place unless open reduction is performed



FIG 122 Fracture of the Patella There is a fracture of the upper third of the patella with marked upward displacement of the proximal fragments The soft tissues of the prepatellar region are markedly thickened and swollen

Fractures of the Lower Leg

Fractures of the tibial spine are rare The fracture is usually not actually of the tibial spine but an avulsion of the attachment of the anterior cruciate ligament with separation of an osteochondral fragment in the region anteriorly to the tibial spine The fragment may be completely separated or be displaced only slightly at its posterior margin The fracture is the result of forcible forward displacement of the tibia in relation to the femur or abrupt inward rotation of the tibia The lateral tibial spine may be fractured without avulsion due to impaction by the lateral femoral condyle

Fractures of the upper end of the tibia have become increasingly frequent in recent years The so called fender or bumper fracture resulting from the impact of an automobile against the leg of a pedestrian causes a very characteristic lesion The fracture is longitudinal or oblique and extends to the articular surface and condyle of the tibia The fragment is displaced downward and outward It appears impacted usually due to overlapping The diagnosis is estab

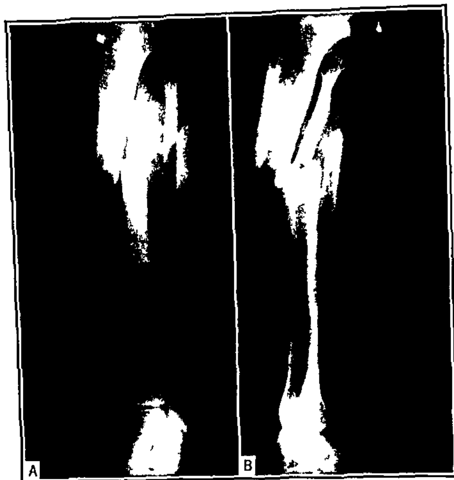


FIG 123 Fractures of Both Bones of the Lower Leg with Separation and Displacement of the Fragments *A* Anteroposterior view *B* Lateral view



FIG 124 Fracture of Both Bones of Lower Leg with Synostosis There are united fractures of the upper third of both bones of the lower leg A large bony bridge extends between the tibia and fibula at the site of the fractures

lished by depression of the lateral condyle of the tibia, widening of the outer half of the knee joint space and the fact that the outer edge of the tibia extends farther laterally than the corresponding edge of the femur It is a serious fracture because of the resultant derangement of the knee joint Prompt reduction is essential Fractures of the spines of the tibia often occur in association with fracture of the condyles The head of the fibula may be fractured simultaneously Fractures of the upper end of the fibula are relatively unimportant as there is little or no disability and healing ensues promptly

The tibia or fibula may be fractured at any portion of the shaft If only one of these bones is involved, reduction is not difficult and healing is satisfactory and prompt Fractures of both bones of the lower leg present a different problem As a rule, the fractures are spiral or oblique Care must be exercised to include the entire leg from ankle to knee on the roentgenogram as the tibia may be broken at its lower end and the fibula at the head If the line of fracture of the tibia is extended by an imaginary line, the site of fracture of the fibula will most probably be found at the point where the extension of the line of the fracture of the tibia crosses the fibula With a transverse fracture of the tibia the fibula is usually broken at the same level as the tibia Apposition of the fragments in fractures of both bones is frequently difficult or impossible and the traumatism attendant on manipulation in attempting to obtain exact anatomical alignment makes this procedure unwise The weight-bearing lines are of the utmost importance and should be re established if possible Shortening of one inch or less is usually not significant and apposition of one-half or more of the fractures surfaces is satisfactory in most instances Callus is slow to form in fractures of both bones of the lower leg and the prognosis must be guarded Weight bearing should be forbidden until bony callus is present Non union is more frequent in fractures of both bones of the lower leg than elsewhere

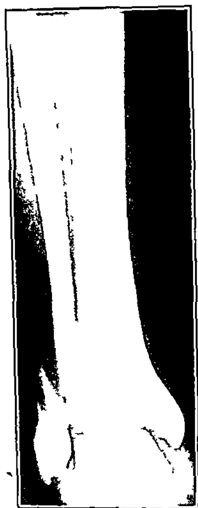
Fracture of the Ankle

Fractures of the lower end of the fibula, the external malleolus, are among the commonest fractures of the human bones The internal malleolus is usually fractured at the same time the so called Pott s fracture Frequently there is also a fracture of the posterior aspect of the lower end of the tibia the line of fracture extending into the articular surface of the tibia There is usually lateral displacement of the foot as well as the fragments of the malleoli and posterior displacement of the astragalus may also occur The reduction must re establish the normal weight bearing lines and the mortise of the ankle Roentgen studies soon after the reduction are essential

Fractures of the Foot

Tarsal Bones Fractures of the calcaneus are among the most frequent and the most important of the tarsal fractures They usually result from a fall from a height with the patient landing on the feet They may occur in conjunction with fractures of the spine pelvis and femur and the more serious injury may result in the calcaneus fracture being overlooked The fractures are frequently comminuted and there is often separation of the fragments Early reduction is important as it usually is impossible to

obtain satisfactory position of the fragments after healing has begun to take place. Anteroposterior views are difficult to obtain and lateral views are not always sufficient. Careful attention to the technical details of the roentgen study is essential and films in various projections are imperative. Callus is very slow in forming. Fibrous union is the rule and it may be many weeks before bony callus is visible. Fractures of the sustentaculum tali are important as they often result in disturbance of the arch of the



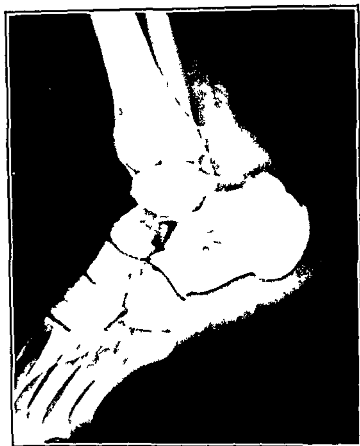


FIG 125 FIG 126

FIG 125 Fracture of Lower End of Fibula. There is a fracture of the lower end of the fibula with lateral displacement of the distal fragment. There is widening of the mortise of the ankle joint with slight medialward displacement of the lower end of the tibia with relation to the talus.

FIG 126 Fracture of Talus

foot. Fractures of the talus and other tarsal bones are frequently chip fractures or periosteal tears. However, severe twisting falls or the passing of a heavy wheel over the foot may cause crushing fractures with severe injury to the arch of the foot. Replacement of the arch is as important as the reduction of the fracture.

Fractures of the Anterior Process of the Calcaneus. There is a form of fracture of the anterior process of the calcaneus which may not be associated with serious complications. In many instances the fracture of the anterior process is considered a sprain. The diagnosis is difficult

and the condition is overlooked unless the possibility of the condition is borne in mind The fracture is usually caused by sudden dorsiflexion of the foot on the ankle The resultant compression of the lateral antero superior portion of the calcaneus by the adjacent cuboid produces the fracture It is commonly industrial in origin, a frequent cause being the striking of the posterior portion of the ankle or calf by a hand truck from behind The fracture may be caused by forcible plantar flexion of the foot or may be the result of the shearing action on the bone as it is rotated



Fig 127 Fracture of the Calcaneus An extensive linear fracture of the calcaneus is clearly visualized The fragments are not displaced



Fig 128 Comminuted Fracture of the Calcaneus and Fracture of the Proximal End of the Fifth Metatarsal The Calcaneus shows a comminuted fracture with separation of the fragments The soft tissues of the heel are very markedly thickened and swollen There is a fracture of the base of the fifth metatarsal with overlapping of the fragments

under traumatic conditions against the cuboid The fracture may consist of a small break at the anterior edge of the bone It frequently is not recognized on the first roentgenograms and is demonstrated only after repeated roentgen study in an effort to explain the painful disability Many of the patients describe their injury as a twist The use of high heeled shoes with plantar flexion and abduction is a contributory factor

In many individuals the anterior process of the calcaneus is poorly developed The lateral half of the floor of the bone may rise sharply upward to produce a well defined triangular beak which is more vulnerable to the effects of avulsive, compressive or torsional forces As the bifurcated ligament is attached to this process sudden adduction of the forepart of the foot at the mediotarsal joint especially in equinus, exerts a strong tension upon the bone and results in fracture In the typical case there is immediate pain along the outer aspect of the mid portion of the foot and discomfort on weight bearing Tenderness is localized to an area about 1 cm in diameter at the dorsal surface of the calcaneocuboid joint at a point about 4 cm anteriorly and slightly below the tip of the external malleolus There is spasm on adduction Swelling at first is limited and then becomes widespread The point of maximum tenderness can be clearly localized and is important in diagnosis

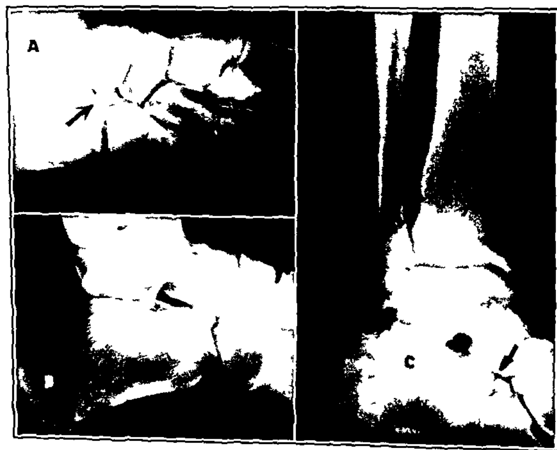


FIG 129 Fracture of the Supero anterior Process of the Calcaneus A Left foot one week after the injury There is a fracture of the supero anterior process of the calcaneus (arrow) with slight separation of the fragments B Lateral projection of the right foot for comparison showing the normal appearance of the supero anterior aspect of the calcaneus C Lateral projection of the left foot eight weeks after the injury The fracture is well shown the fracture line being more clearl defined than in the study immediately after the injury (arrow) The fragment is ununited and is displaced downward

In order to establish the diagnosis, properly made roentgenograms are of the utmost importance. The routine anteroposterior view of the foot is seldom helpful and a direct lateral is usually not sufficient to demonstrate the fracture, particularly if the fracture line casts only a faint shadow or there is no appreciable displacement of the fragments. Oblique views at various angles are essential to separate the shadow of the anterior process from that of the talus. A magnifying glass should be utilized in the study of the roentgenogram and the entire area examined very carefully. Unless suspected and carefully sought for, fractures at this site often remain undiagnosed. Since the fractures are relatively rare, they are frequently overlooked. The presence of a trigger point of tenderness gives a clue to the diagnosis and stresses the necessity of careful and complete roentgen study.

Avascularization of the Talus. The talus is composed of a body, head and neck. The body articulates with the tibia superiorly, the fibula laterally and the calcaneus inferiorly. The head articulates with the navicular and the calcaneus. The neck comprises the constricted portion of the bone between the body and the head. The bone receives its blood supply from a branch of the dorsalis pedis artery which enters the lateral aspect of the neck and a few small tributaries which enter at the ligamentous and capsular attachments. Fractures of the talus usually involve the neck. A shearing force tends to fracture the neck transversely. Fractures proximal to the point of entry of the dorsalis pedis artery affect the blood supply and predispose to avascularization of the proximal fragment. Following a fracture, there may be generalized decalcification and osteoporosis of all the bony structure in the region of the fracture. The extent of the decalcification is dependent upon the degree of interference with the blood supply to the bone. Decalcification affects only the segments which are adequately nourished, the portion of the bone which is not supplied with blood remaining of normal density. In fracture of the neck of the talus proximal to the dorsalis pedis artery with resultant avascularization of the proximal fragment, roentgen study shows marked contrast between the normal density of the avascular fragment and the pronounced decalcification of the surrounding bones. This may persist for many months and is of great significance in that the period of immobilization may have to be prolonged until union and revascularization have taken place.

Metatarsals. Fractures of the metatarsals are usually due to heavy objects falling on the foot or a wheel passing over the foot. Fractures of the proximal end of the fifth metatarsal usually result from a fall or twist of the ankle and attention may be directed to the malleoli rather than to the foot. It is good practice to include the proximal portion of the foot in all ankle films. Displacement in metatarsal fractures is difficult to reduce. The arches of the foot must be maintained. Numerous sesamoid bones are commonly present and must not be confused with fractures. Studies of both feet must be made to assist in identification of the sesamoids.

Phalanges of the Feet. The phalanges are usually fractured by stubbing of the toe, dropping a heavy object on the toe, or the passing of a wheel over the foot. Comminution usually results. Involvement of the articular surfaces is frequent. Healing is usually prompt. Fractures of the great toe are very disabling because of weight bearing. Two sesamoids are usually present in the great toe and may be misinterpreted as fractures by the uninitiated.

Sesamoids The anatomical location of the sesamoid bones makes them particularly liable to trauma Multiple ossification centers are of frequent occurrence and these may unite or remain permanently ununited, failure of union being termed congenital division Bipartition, tripartition, and quadripartition may result It is of the utmost importance that congenital partition be differentiated from fracture from the surgical and medico legal points of view Fractures of these bones are rare Fractures of the sesamoids of the great toe, thumb, tibia, and fibula have been reported and the progress of the healing has been demonstrated by roentgen studies The fracture may result from direct or indirect trauma Associated fractures of the adjacent bones are frequently present, particularly after a crushing type of injury Differentiation from a partite sesamoid may be difficult Fractures are more common in women who wear high heeled shoes and whose occupation entails prolonged standing Roentgen study is essential in all cases of suspected fracture The examination in the case of the pedal sesamoids is best made with the toe in dorsiflexion Anteroposterior and lateral views are essential Both sides should be examined as developmental variations are frequently bilaterally symmetrical The criteria for the establishment of a diagnosis of fracture comprise the following (1) an irregular, serrated fracture line or lines (2) separation of the fragments, (3) the presence of callus or other evidences of healing, (4) absence of similar changes in the contralateral sesamoid bone Immobilization and rest usually suffice to bring about a cure Removal of the fragments is rarely necessary

SKULL FRACTURES

Fractures of the skull have shown a very marked increase in frequency during recent years due principally to automobile and industrial accidents In some cases they may occur in association with little *or no* trauma to the brain, while in others there is severe internal injury which may result fatally In the majority however, there is associated brain injury in only about 6 per cent is no damage to the brain manifest Although fracture of the skull is in every instance a matter of concern the extent of injury to the brain is of much greater consequence in evaluating the danger to the patient than is the damage to the bones Depressed and compound fractures are particularly important They are almost invariably associated with brain damage and the clinician and neurosurgeon must know the condition of the bone as exactly as possible in order to plan proper treatment With extradural hemorrhage there is practically always a fracture and its precise location is of great importance in treatment Fractures of the skull are classified into numerous subdivisions the most common of which are linear, depressed, stellate, and diastasic They may involve the vault, the base or both and may be simple or compound, the latter comprising those which underlie a laceration of the soft tissues or traverse the cribriform plate the nasal accessory sinuses or the mastoid A diastasic fracture is one involving separation of the bones at a suture line

Roentgenography offers an accurate dependable, and simple method of determining the presence and type or absence of fracture It can usually be carried out without pain or discomfort and is essential in all suspected cases It is of greater value than any or all other methods of examination

and is properly used in conjunction with but not to the exclusion of other clinical studies The roentgenograms reveal the extent, location, and nature of the injury to the bone often help in the choice of the measures required for treatment, and serve to reveal the presence of underlying bone disease such as malignancy, osteomyelitis, and similar complications They are also of value in supplying permanent records of the damage to the bone that can be referred to in studying the progress of the case

There has been considerable controversy in the literature as to the time when roentgen studies of the skull in cases of suspected fracture should be made If the patient s condition permits the roentgenograms should be taken soon after the trauma In those who are comatose irrational, or in severe shock the examination may well be delayed until improvement occurs A complete study is best made with a cooperative individual who can assume the necessary positions and lie quietly during the exposures



Fig 130 Fracture of the Skull There is an extensive linear fracture of the posterior parietal bone and the temporal bone The fracture line is widest in its superoposterior portion becoming narrow and faint in the temporal region It crosses several blood vessel grooves

Turning of the head and manipulation are essential to a complete roentgen study and these or attempts at restraint of irrational struggling patients may do harm Portable roentgenograms are seldom satisfactory as the small bedside type of machine requires a long exposure time which not infrequently results in films lacking in contrast and detail particularly if the Potter Bucky grid cannot be used However there are times when roentgen studies which are incomplete and not entirely perfect because of having been made soon after the injury and with the patients who are uncooperative or in severe shock may afford very valuable data Thus if a patient is thought clinically to have a subdural hematoma simple sagittal views may show pineal displacement and therefore be of great importance in determining the location and possibly also the extent of the bleeding In compound or depressed fractures roentgen studies afford an indication of the severity of the fracture and help in planning the surgical approach or method of therapy With proper selection of the case by the neurosurgeon and close cooperation between the clinician and radiologist much may be accomplished in cases of these types

Roentgen Technic. The technic of the roentgen examination of the skull in cases of suspected fracture is of the utmost importance. Films must be taken in the anteroposterior, posteroanterior, and both lateral projections with stereoscopic views on the side of the trauma whenever possible. In addition, occipital (Towne) views are made routinely in the clinic with which I am associated. Tangential projections through the site of the trauma may be helpful in showing depression, comminution,



Fig. 131. Depressed Fracture of the Skull. There is a comminuted fracture of the frontal bone with a large defect and increased density at the margins of the depression. *This type of fracture results from a blow by a blunt instrument such as a hammer.*



Fig. 132. Fracture of the Left Temporoparietal Region

or separation of fragments Orbital views are used if indicated In cases with bleeding from the ear the mastoid areas are studied Basal fractures are among the most difficult to demonstrate Bleeding from the nose, mouth, or throat after an injury may result from a fracture in the base and is an indication for additional studies, particularly submento vertical and vertico submental projections and special views of the occiput and foramen magnum Each roentgenogram of the skull must be of perfect detail and definition as very slight motion or lack of clarity of outline may result in failure to demonstrate a fracture Every film must be studied very carefully as only a single film of the entire series may reveal the break in continuity and prove the presence of a fracture One should not hesitate to resort to re examination if the roentgenograms are not of perfect quality or if further study appears indicated for any reason whatever Dressings may obscure a fracture and should be re moved when possible If the surgeon feels it is inadvisable to disturb dressings, re examination may be necessary at a later date

It must be borne in mind that a very severe trauma may not result in a fracture while slight blows can cause very extensive fractures Not in frequently, the roentgenologist is requested to study only the site of the trauma This is always unwise as fractures may occur at distant points The so called contrecoup fractures are on the side opposite the injury In cases of gunshot wound the roentgenologist may be called on to localize and help remove foreign bodies and bony fragments This requires stereoscopic studies and, in some instances roentgenoscopic observations Similarly compound fractures at times present imbedded foreign bodies and depressed fractures may have displaced fragments which can be localized accurately Infants and young children always pose a special problem as they offer particularly great difficulties in immobilization and proper positioning Patience and persistence are essential for obtaining perfect roentgenograms and repeated attempts must be made to obtain the necessary views

Roentgen Findings In the roentgenogram with a linear type of fracture of the skull, there is a sharply outlined clear cut linear shadow which represents the break in the continuity of the calvaria The so called "bevel fracture may involve the inner and outer tables at slightly differ ent levels the obliquity of the x ray beam causing two distinct lines to be visible on the roentgenogram Fractures in the temporoparietal region may be demonstrable only on the lateral projections the sagittal views not revealing any abnormalities Similarly occipital fractures may be seen only in the Towne position The side on which the fracture is located may be difficult to determine at times but this can usually be done with a reasonable degree of accuracy if one bears in mind that the fracture is more clearly defined and appears smaller when it is on the side of the head nearer the film There are many instances in which the sagittal projec tions do not show the fracture and it may at times be impossible to obtain stereoscopic views because of the patient s condition Fractures may encircle the skull and be visible on both sides Here, the extent of in volvement of each side may be determined by the above described methods or the use of stereoscopic views In depressed fractures there is an irregular area of increased density at the site of the depression As with other flat bones the fracture line may be more clearly visible after an interval of a few weeks owing to slight absorption along the line of fracture Also, the soft tissue swelling and edema are usually less marked

at this time than immediately after the trauma, making the fracture line more easily demonstrable

Roentgenoscopy alone cannot be relied on for the diagnosis of skull fracture A thorough knowledge of the anatomy of the skull is essential as suture lines blood vessel grooves, diploic channels, and certain congenital or developmental anomalies may closely simulate fracture Only a competent roentgenologist should attempt a diagnosis in cases of suspected skull fracture Anatomic ridges have frequently been mistaken for fracture by the inexperienced observer Stellate venous channels may be confusing, and this also is true of emissary veins, Pacchionian depressions dural or diploic venous lakes, and sutures, particularly metopic or aberrant suture lines The inner aspect of a suture may at times be seen as a straight, smooth line and has been mistakenly diagnosed as a diastasic fracture The arterial grooves usually present somewhat symmetrical patterns on the two sides, become progressively smaller toward the vault, bifurcate regularly, and are smoother and less sharply defined than fracture lines Fracture may occur within or across vessel grooves and this should be indicated in the roentgen report as it is of importance in therapy and prognosis If the fracture communicates with an air sinus special note of this fact and of any abnormal density of the sinus should be made

In infants and young children the unossified bones must be differentiated from fractures Wide separation or displacement of fragments may be mistaken for suture lines Neonatal fractures are usually of the linear or fissure type and appear as lines of increased radiance often with wide separation The comminuted stellate type of fracture with depression which occurs usually with the application of forceps during delivery is of frequent occurrence Depression and overlapping of the calvaria may result after long, difficult labors due to pressure of the head against the sacrum or symphysis The parietal and frontal bones are most commonly involved No special therapy is required as the bones return to normal position spontaneously in practically all cases

Old Fractures Old fractures are characterized by thin, smooth margins While simple linear fractures may fill in and disappear in a few months or one to two years many skull fractures heal only by a fibrous line and may not calcify for long periods if at all Leptomeningeal cysts may form in association with old fractures

Glaser and Blaine reviewed a series of 150 cases in studying the fate of cranial defects secondary to fracture and surgery Their conclusions are briefly summarized as follows Linear fractures in children under six years of age usually disappear in six to twelve months If there is wide separation of the fragments however, the defect may persist for long periods of time and in some instances is permanent In adults, linear fractures begin to fade soon, usually disappearing partially in seven months and completely in two to four years These figures apply to fractures of the frontal, parietal and temporal bones In the occiput, healing takes much longer, at times requiring as long as eight years In some cases there is absorption along the margins of the fracture, leaving a permanent defect of variable size This may occur in both children and adults and is known as post traumatic fibrosis Depressed fractures with no associated elevation of fragments become rounded and smooth the fracture lines are not demonstrable, although the depression may persist permanently

Operative Defects of the Skull

Operative defects of the bones of the skull consist of trephine openings, bone flaps, or areas from which bone has been removed surgically. Trephine holes are recognized by the fact that they are small, rounded, and present sharply outlined margins. They are usually in the posterior parietal regions and there is no irregularity of outline or bone proliferation about the edges of the area. Small trephine openings may be made asymmetrically and vary in size and location. A knowledge of the previous history is essential for proper diagnosis. Operative defects are usually recognized without difficulty. Wide defects in adults are permanent and do not decrease in size although the margins of the area tend to become smoother and rounded. In children, defects of considerable size may decrease markedly in area with the passage of time. Bone fragments *in situ* postoperatively show spaces between the fragments unless very closely approximated. Bone flaps may undergo partial absorption or remain essentially normal in appearance. Osteoperiosteal flaps usually do not become united by osseous union for long periods and may not be definitely reunited even after many years. Bone chips are clearly visible in operative defects and present a characteristic appearance.

Metallic plates are frequently used to cover operative defects and are left *in situ* permanently. Tantalum and vitallium are the metals most commonly used as they are firm and noncorrosive. Other cranioplastic materials such as acrylic resin and celluloid are not opaque to the roentgen ray and therefore do not interfere with later x-ray study. Metallic brain clips and wires employed to secure osteoperiosteal flaps are easily recognized by the experienced roentgenologist. The rubber or plastic tubing employed in the Torkeldsen or other short circuiting procedures for internal hydrocephalus may be visualized in the roentgenogram.

Fracture of the Nasal Bones

Fractures of the nasal bones are easily demonstrated if carefully sought for. They are not well shown in routine skull films, lateral views with special exposures being essential. A small dental film pressed against the side of the nose, studies in the Waters position, and bite films held in the mouth are helpful. Fractures of the nasal bones usually are transverse in type. They may be oblique or longitudinal and must be differentiated from the depressions for the nasal nerve branches. The site and extent of the fracture and displacements can be established by roentgen study. Dislocation of the naso frontal, naso maxillary and naso nasal sutures should be observed carefully. Associated fracture of the nasal spine of the maxilla occurs frequently. Early accurate diagnosis with immediate reduction and replacement of the fragments is essential. If the fractures unite in malposition, refracture is necessary for correction of deformities. In children, a slight deformity may become more noticeable as the child grows older.

Fracture of Zygomatic (Malar) Bones

The zygomatic bones form the external wall of the orbital cavity and the prominence of the cheek. Fractures in this region are very important because of possible disfigurement. The fracture may be obscured soon after the trauma because of associated soft tissue swelling and hemor-

rhage After the swelling disappears, displacement of the fragments becomes prominent and may be difficult to correct because union has begun The most common site is at the zygomatic aspect of the zygomatico-temporal suture The fragments are usually displaced and there is flattening and depression of the cheek

Fracture of the Maxilla

Fractures of the maxilla are very important because of associated involvement of the orbit, mouth, nose, sinuses, alveolar processes and teeth The fracture is usually the result of crushing injury of the face and is often associated with fracture of the skull It is necessary to demonstrate the lines of fracture, fluid in the sinuses, and dental fractures and displacements



Fig 133 Fracture of the Right Zygoma There is a comminuted fracture of the right maxilla and zygoma with separation of the fragments There is markedly increased density over the right antrum due to the presence of blood in the sinus



Fig 134 Fracture of the Maxilla and Zygoma There is a markedly comminuted fracture of the left maxilla and zygoma with wide separation of the fragments The fracture involves the orbit and the antrum The left zygomatico-frontal suture is separated The left eye was enucleated and a prosthesis inserted manifested by the ring like shadow in the left orbital region

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Fig 135 Fracture of the Mandible There is a linear fracture of the body of the mandible The line of fracture extends along the margin of the inferior bicuspid There is separation of the fragments

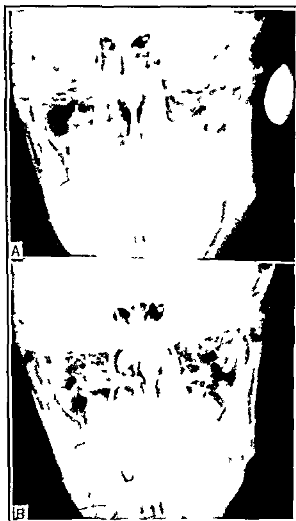


Fig 136 Fracture of the Mandible *A* There is a fracture of the mandible in the region of the junction of the body and the ramus with separation of the fragments The roentgen study was made six hours after the injury *B* Four months later The fracture line is partially filled in The fragments are in good apposition and alignment

Fracture of the Mandible

The mandible is the most frequently fractured bone in the face. It usually is broken in two places, often on opposite sides. The fracture may result from direct trauma or during tooth extraction. It is necessary to take multiple views to show the various portions of the bone. Fractures are most common at the angle and in the canine fossa. The proximal fragments are usually displaced upward and medially. Fracture of the condylar process requires special lateral views and may be shown on skull or mastoid studies. Fractures or displacements of teeth are important. Associated abscesses of the teeth or compound fracture are apt to be complicated by osteomyelitis. Delayed or non-union is common and may be due to many causes, the chief of which are infection, fractured teeth, poor reduction with interposition of soft tissues, loss of bone as in a gunshot wound or compound fracture, lues, tuberculosis, lack of immobilization, and pathologic fracture in metastatic neoplasm or cysts.

SPINE FRACTURES

Fractures of the Cervical Spine

Fractures of the cervical spine occur as the result of diving into shallow water, striking the head on the top of an automobile, or a fall. Great care must be exercised not to increase the extent of the injury by manipulation of the patient. In fractures of the upper cervical vertebræ, sudden hyperextension may result in paralysis or death. The head must be supported and only such roentgenograms made as may safely be taken at the time. The lateral view can be made with the patient supine and the film resting against the shoulder. Fracture of the first cervical vertebra is difficult to demonstrate as the details are obscured in the usual views. Semi lateral or oblique projections are required and tilting the head to the side may bring the fracture into view. Fracture of the dens is relatively common and is not infrequently associated with dislocation. In complete fracture of the dens, the upper fragment and the head are displaced anteriorly and in the lateral projection the posterior aspect of the first cervical vertebra lies anteriorly to the tubercle of the axis. Posterior displacement is rare. The fracture most commonly involves the base of the dens. The fracture may be visualized more clearly after the lapse of a few days or weeks as absorption develops along the line of fracture and re examination is essential in all suspected cases. Fractures of the middle and lower cervical vertebræ are usually of the compression type and may be comminuted. Associated dislocation is frequent. The affected body shows wedging with narrowing of its anterior portion. Both the superior and inferior surfaces are usually affected. Fractures of the articular facets may occur alone or in association with fractures of the body or lamina.

Fractures of the Spinous Processes

Spinous process fractures are rare. Their recognition is important as they are frequently overlooked. The lateral view is essential for demonstration of these fractures. On the anteroposterior roentgenogram they



FIG 137 Dislocation of Sixth Cervical Vertebra with Fracture



FIG 138 FIG 139

FIG 138 Healed Fracture of the Cervical Spine There is an old compression fracture of the fifth cervical vertebra There is bony bridging between the anterior aspects of the fourth to sixth cervical vertebræ The fracture was sustained eighteen months previously

FIG 139 Compression Fractures of the Fourth and Fifth Dorsal Vertebræ The bodies of the fourth and fifth dorsal vertebræ are narrowed and irregular in outline There is a defect in the supero anterior aspect of the body of the fifth dorsal vertebra and the fourth vertebra projects into this defect The patient was a paratrooper whose parachute failed to open completely He dropped 900 feet landed on his feet and fell forward in a jackknife position

are recognized only with difficulty. Fractures of the spinous process occur most commonly in the lower cervical and upper thoracic vertebræ and only occasionally in the lumbar region. In the anteroposterior roentgenogram the fracture is manifested by malalignment and downward displacement of the distal portion of the spinous process. A double shadow is demonstrable in the region of the spinous process. A smooth, crescentic area of increased radiance represents the fractured base of the spinous process and a shadow of increased bony density in the soft tissues slightly caudad to the base of the spinous process represents the displaced tip. Patients with a severe injury to the cervicodorsal region usually maintain the head in hyperflexion or hyperextension. On the lateral projection of the neck, the spinous processes of the seventh cervical and first thoracic vertebræ may be completely obscured because of superimposition of the shadows produced by the bones and soft tissues of the shoulder girdle. In this instance, the anteroposterior view is essential to establish the diagnosis. In fractures of this type, careful search must also be made for associated fractures of the bodies of the cervical or thoracic vertebræ. The radiographic manifestations which permit of identification of fractures of the spinous processes of the lower cervical and upper thoracic vertebræ have been stressed by Zanca and Lodmell. The associated soft tissue swelling and thickening make the fracture difficult to demonstrate and in the past it has been unrecognized in many instances.

Lumbar and Dorsal Spine Fractures

Fractures of the dorsal and lumbar vertebra may be of the linear or compression type. In the latter group, there is wedge formation with narrowing of the anterior aspect of the body. The body is widened in its anteroposterior diameter. The fracture usually results from jackknifing or hyperflexion of the spine. The associated dislocation is of importance in establishing the degree of traumatism to the cord. The compression may affect only the superior or inferior aspect of the body of the vertebra or one lateral margin although this is less frequent. Healing of fractures of the spine is usually by fibrous union with gradual disappearance of the fracture line and the development of increased density at the site of the fracture. After the lapse of a period of months new bone may be formed along the anterior aspect of the vertebral body with a bony bridge extending from the affected vertebra to those above and/or below. In some instances there is no evidence of new bone formation at any time.

Fractures of the laminæ may be demonstrable only on stereoscopic, oblique or other special views. Fractures of the spinous processes may be easily overlooked, particularly in the anteroposterior view. In the lateral view, the fracture may be blotted out by overexposure. Large amounts of callus may result in the presence of wide separation of the fragments. Therefore early recognition and prompt therapy are important. Fractures of the transverse process of the lumbar vertebræ are frequent and may result from direct trauma or muscle pull such as occurs after a misstep or slipping on the curb or a stair. The line of fracture may not become visible for days or weeks. Separation of the fragments is not uncommon. There is bulging or obliteration of the shadow of the psoas muscle. Large callus formation may ensue.

Fractures of the Vertebræ in Convulsive Shock Therapy

The first report of a convulsion fracture appeared in the literature in 1938 and since then the condition has become recognized as being of relatively frequent occurrence There has been a definite decrease in the number of cases in recent years One of the most important factors in this regard is the utilization of electric shock to replace the older forms of therapy With metrazol, the agent originally utilized for the induction of shock, there were relatively large numbers of fractures, in some series



Fig 140



Fig 141

Fig 140 Old Fracture of the Body of the Eleventh Dorsal Vertebra The body of the eleventh dorsal vertebra is wedged irregular in outline and eburnated There is bony bridging along the anterior margins of the tenth eleventh and twelfth dorsal vertebræ indicating that the fracture is of long standing There is a moderate kyphosis in the dorsolumbar region

Fig 141 Compression Fracture of the Eighth Dorsal Vertebra after Shock Therapy The patient had received electroshock therapy The fracture resulted during the treatment The affected vertebra is wedged anteriorly and its superior surface is depressed eburnated and irregular in outline

as high as 47 per cent Electroshock is the method now in common use and fractures develop in less than 5 per cent of the patients receiving the treatment Huddleson and Gordon made a survey of 252 patients for anomalies of the spine which might predispose to fracture after electric shock therapy The abnormalities comprised rarefaction of the vertebral bodies decreased density of the vertebral discs narrowness and haziness of the intervertebral spaces atrophic and hypertrophic arthritis abnormal curvatures and similar conditions They noted an over all fracture incidence of 6 3 per cent with 4 3 per cent in normal spines and 6 8 per cent in abnormal spines It appeared that the abnormalities demon

strated on a pre shock film had no apparent significance in relation to the subsequent incidence of fractures.

The use of curare results in a definite lowering of the incidence of fracture. Curare does not appear to be of value in arresting the progress of the fracture or preventing the appearance of new fractures once the treatment course has begun and evidence of injury to the spine has been discovered. The use of the drug entails a definite risk and it should be limited to selected cases. The presence of vertebral osteochondrosis appears to impart a resistance to the patient's spine to electric shock fracture, the incidence of fracture in these cases being very low. Other factors such as race, deforming spondylosis, kyphosis, Schmorl's nodes, and the physician who administers the shock appear to have no relation to the incidence of convulsion fractures. The significant factors in the etiology are the duration, direction, and intensity of the forces applied to the spinal column. The number of convulsions appears less significant in this regard. The dorsal spine between the levels of the third and eighth vertebræ is most liable to suffer fractures because of the relatively limited degree of motion and the powerful musculature in this region. During the convulsion there is partial flexion in the clonic phase, and this is considered as being the causative factor in the production of the fracture. In the tonic phase, the spine becomes hyperextended and Webb is of the opinion that the fractures ensue at this time because the muscular contractions reach the maximum. It is believed that the injury constitutes a disc repercussion fracture and that rapidly repeated hammering of the disc surfaces in the least mobile portion of the spine results in compression of the superior aspects of the vertebral bodies. The incidence of fractures is highest in males, probably because of the stronger musculature. Persons thirty to thirty nine years of age are most prone to suffer fractures of the spine after electroshock therapy. The fracture may involve a solitary vertebra, although in most instances several bodies are affected. The cervical spine has not been involved in any of the reported cases. A single body may escape injury while others immediately above and below have suffered fractures.

In the great majority of cases, the fracture is manifested roentgenographically by a slight depression of the middle portion of the superior surface and a narrow band of increased density within the vertebral body slightly below the surface due to impaction of the spongy bone. No fracture line may be demonstrable in the early stages. The sagittal projection in many instances does not reveal any abnormality in the spine, the lateral view being essential for diagnosis. In the presence of associated herniation of the nucleus pulposus, there is narrowing of the intervertebral space. This manifestation occurs more commonly in the convulsion fracture of shock therapy than in other types of compression fractures of the vertebræ. More severe types of fractures result in compression of the anterior aspect of the vertebral bodies with varying degrees of wedging. The posterior aspect of the vertebral body is seldom involved. Fracture lines may be present and usually involve the superior articular surface. There may be marked narrowing, wedging, and elongation of the affected vertebræ. The inferior surfaces of the vertebral bodies may be involved, although this is less common. The fractures seldom constitute serious complications and do not as a rule eventuate in arthritis, permanent deformity, or serious disability. Kyphosis usually does not ensue. This is in striking contrast to tetanus, in which practically all cases develop a

Fractures of the Vertebræ in Convulsive Shock Therapy

The first report of a convulsion fracture appeared in the literature in 1938 and since then the condition has become recognized as being of relatively frequent occurrence There has been a definite decrease in the number of cases in recent years One of the most important factors in this regard is the utilization of electric shock to replace the older forms of therapy With metrazol, the agent originally utilized for the induction of shock, there were relatively large numbers of fractures, in some series





Fig 140 Fig 141

Fig 140 Old Fracture of the Body of the Eleventh Dorsal Vertebra The body of the eleventh dorsal vertebra is wedged irregular in outline and eburnated There is bony bridging along the anterior margins of the tenth eleventh and twelfth dorsal vertebræ indicating that the fracture is of long standing There is a moderate kyphosis in the dorsolumbar region

Fig 141 Compression Fracture of the Eighth Dorsal Vertebra after Shock Therapy The patient had received electroshock therapy The fracture resulted during the treatment The affected vertebra is wedged anteriorly and its superior surface is depressed eburnated and irregular in outline

as high as 47 per cent Electroshock is the method now in common use and fractures develop in less than 5 per cent of the patients receiving the treatment Huddleson and Gordon made a survey of 252 patients for anomalies of the spine which might predispose to fracture after electric shock therapy The abnormalities comprised rarefaction of the vertebral bodies decreased density of the vertebral discs narrowness and haziness of the intervertebral spaces atrophic and hypertrophic arthritis abnormal curvatures and similar conditions They noted an over all fracture incidence of 6 3 per cent with 4 3 per cent in normal spines and 6 8 per cent in abnormal spines It appeared that the abnormalities demon

INTRAUTERINE FRACTURES

Intrauterine fractures may occur as a result of trauma with direct injury to the fetus, amniotic shearing, and congenital lesions such as osteogenesis imperfecta, syphilis, and chondrodystrophy. Obstetrical fractures are ruled out by roentgenograms made on the first or second day of life. If repair of a bone fracture is evident at this time, the diagnosis of intrauterine fracture is established with definiteness. The lesions most commonly affect the tibia and fibula, although the bones of the arm may also be involved. Injury to the long bones is common, particularly in breech deliveries. The severe torsion of a breech delivery may cause the bones of the lower extremity to be fractured at the weakest point, the epiphysis. The periosteum is stripped off and hemorrhage may occur.

MARCH FRACTURE (FATIGUE FRACTURE)

March fracture was originally described by Breighaup in 1855. The first roentgenogram of this condition is believed to have been taken by Stechow in 1897. The term is applied to fractures which occur without great trauma, most often during a march. The condition has been reported in the metatarsals, the tibia, the femoral neck, the supracondylar area of the femur, and other bones. The concept of fatigue fracture of

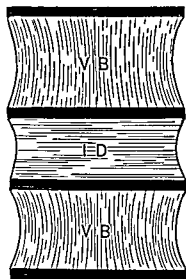




Fig 142. Herniation of the Nucleus Pulposus (Schmorl's Disease). *A*, Diagram showing the normal nucleus pulposus. VB = Vertebral Body; ID = Intervertebral Disc. *B*, Schmorl's Nodule. There is a rounded area of increased radiance involving the superior aspect of the body of the second lumbar vertebra, the characteristic manifestation of herniation of the nucleus pulposus into the vertebral body.

spinal deformity similar to that in juvenile kyphosis whether vertebral fracture was present or not Prompt and complete healing results with the usual type of callus formation Progressive wedging does not ensue unless the vertebræ are pathological

Shock therapy has been utilized in patients with osteoarthritis of the spine, varying degrees of porosis, scoliosis, kyphosis, and other anomalies A history of a previous fracture does not constitute a contraindication to shock therapy as new fractures or refractures of previously fractured vertebræ have been recorded without untoward results Healing takes place satisfactorily and without complications or sequelæ Additional pathologic changes do not develop in the affected vertebræ to any greater degree than under otherwise normal conditions The complication of vertebral fracture in convulsion shock therapy is not serious and does not require specific orthopedic treatment Intervertebral disc space narrowing is more frequent in the convulsion type of fracture than in the usual compression fracture after trauma suggesting that the mechanism of the former is largely disc repercussion, while that of the latter is primarily hyperflexion

Fractures of the Sacrum and Coccyx

Fractures of the sacrum and coccyx are relatively rare but are very important because of the associated pain and discomfort These fractures are frequently overlooked both clinically and roentgenologically While most commonly caused by a fall they also occur in football players who have been trod upon The break is usually horizontal, oblique or longitudinal fractures being rare Separation of the sacro coccygeal junction is common As with the other flat bones fractures in these bones may be demonstrated better after the lapse of a few days or weeks because of the absorption which takes place Involvement of the sacral foramina is important as indicative of possible injury to the sacral nerves

Injuries to the Nucleus Pulposus (Schmorl's Disease)

The lesion was first described by Schmorl and consists of displacement of the nucleus pulposus from its normal site It may be the result of a single severe trauma or multiple repeated slight injuries Fracture of the cartilage and adjacent bone may occur in association with rupture of the capsule of the nucleus The condition is manifested roentgenographically by a shallow rounded defect measuring 3 to 5 mm in depth and 5 to 10 mm in width in the body of the vertebra and may involve the superior surface the inferior aspect or both There may be narrowing of the intervertebral space with irregularity of the adjacent portions of the bodies of the vertebræ If the nucleus is displaced anteriorly the posterior aspect of the intervertebral space shows narrowing and vice versa Calcification of the nucleus is occasionally seen and is manifested as a rounded slightly irregular dense shadow in the space between the vertebræ

The nucleus is a rounded fluid structure which acts as a shock absorber The forces and shocks are transmitted from one vertebra to the next through the nucleus pulposus With weakening or rupture of the fibrous capsule of the nucleus pulposus the contents herniate Posterior displacement of the nucleus pulposus causes pressure on the cord with resultant pain and reflex changes Lateral herniations may affect the nerve roots with similar manifestations

be of sharp detail A roentgenogram without overexposure of the bone is most satisfactory

March Fracture of the Foot Inadequacy of the feet, abnormal length of the metatarsals overload, neurogenic influences, inflammatory processes and a previously sedentary occupation have been considered possible causes While commonly unilateral it may be bilateral in rare instances In the case of the metatarsals the fracture most commonly involves the middle and distal thirds of the bone the most frequent site being the right third metatarsal The location of sesamoid bones appears to play no part



Fig 143 Fatigue (March)Fracture *A* Anteroposterior view *B* Lateral view There is an incomplete transverse fracture of the upper third of the diaphysis of the tibia The fracture line is partially filled in and there is callus formation about the margins of the fracture

in the production of the fracture Similarly the age, previous occupation neurological disturbances and the anatomy of the feet do not act as contributory factors Muscular fatigue is the single most important factor in the causation of this condition Loss of the resiliency of the muscles permits unusual strain to be thrown on the affected bones and predisposes to or causes the fracture

March Fracture of the Tibia The tibia is second only to the metatarsals as the most common site of occurrence of this type of fracture The usual location is along the medial aspect of the diaphysis at the junction of the middle and upper thirds of the bone It may also occur in the middle third and other portions of the bone The fracture line varies from a faintly visible irregular line of decreased density extending only a short

bone is similar to that implied by the same term as applied to other physical substances A material which is basically sound appears to undergo mechanical dissolution in response to frequent repetition or prolonged concentration of strain on a localized area The fatigue fracture of bone is considered to be the result of repeated slight traumas Underlying systemic disease may play a part in the production of the lesion It has been postulated that the fracture develops as the result of a single injury which for some reason was not recognized by the patient The possibility of an associated underlying infection has been considered Neoplasm formation does not appear to enter into the causation of these fractures Donald and Fitts made a comparative study of two different groups of soldiers one with conditioning, the other with comparatively little conditioning for extensive marches In the well conditioned group only two march fractures were found while in the poorly prepared soldiers sixty cases occured Proper conditioning to marching, together with structurally strong feet and ankles, appeared to be the chief factor in the prevention of march fractures In the German army it was attributed to the upward swing of the leg with outstretched knee and recurving of the supporting leg during rigid marching Pseudofractures of the upper third of the tibia with no history of trauma are common in children from four to sixteen years of age In some cases the fracture is bilateral March fractures of the tibia occur not because of mechanical weakness or anatomical variation of the foot or extremity, but as the result of physiological weakness secondary to fatigue that is brought on by increasing the load which the soldier must carry and by the individual continuing after he tires As the fatigue increases the soldier marches not with his muscle in tone but relaxed and tired Mechanically, the weight is then borne by the bones and ligaments In non military personnel the incidence of march fracture of the tibia is five times as great in the male as in the female The symptoms as a rule are present for several days before roentgen examination is requested The findings on physical examination are characteristic and consist of deep localized tenderness, edema over the site of the lesion, and a bony swelling as callus develops

The lesions have been designated as march shoveler s creeping exhaustion insufficiency overload and spontaneous fracture The most satisfactory name is fatigue fracture The term suggests not a sudden process but one which is gradual and associated with the passage of time 'Pseudofracture is incorrect as these are associated with generalized metabolic disease which influences the metabolism of the skeleton Spontaneous fracture should not be used as this is reserved for the asymptomatic fracture without displacement Similarly the term insufficiency fracture indicates a fracture due to insufficient functional capacity of the bone to meet the stresses placed upon it, which is not the case They should not be classified as pathological fractures because there is no evidence of hyperparathyroidism osteomyelitis rickets osteomalacia or similar underlying disease in the bone The fractures tend to occur at the greatest oscillatory amplitude of a bending bone It may extend transversely or downward and backward Healing usually takes place normally and without sequelæ Diffuse thickening of the cortex in the affected area may persist for several months A small proportion of fatigue fractures cannot be diagnosed on the first examination but become apparent on re examination after an interval of a few days In the demonstration of this lesion it is imperative that the roentgenogram

discovered on examination for a fresh injury or incidentally in examination for other conditions Fractures of the carpal navicular, ribs and transverse processes of the lumbar vertebra are particularly apt to fall in this category In every case of injury of the wrist it is essential that repeated roentgen studies at intervals of several weeks be performed in order to determine whether a fracture which is delayed in presenting roentgen manifestations is present as many so called "sprains of the wrist" are in reality fractures

Unrecognized Skeletal Trauma in Infants Unrecognized skeletal trauma in infants is relatively frequent and in many instances the nature of the lesion is determined only in retrospect on the basis of the roentgen demonstration of irregular fragmentation of the one or more metaphyses



FIG 144 Delayed Appearance of Fracture of the Transverse Processes of the Lumbar Vertebræ *A* Anteroposterior view of the lumbar spine two days after the injury There is no evidence of fracture *B* Seven weeks later There is a linear fracture of the left transverse process of the third lumbar vertebra and a linear fracture of the left transverse process of the fourth lumbar vertebra with extensive callus formation about the margins of the fractures (arrows)

of the tubular bones associated with callus formation manifested by new bone formation external to the shaft Frequently the changes are discovered during roentgen examinations for causes other than known physical trauma In many instances the condition is erroneously diagnosed as scurvy lues developmental anomaly or other serious disease with serious prognostic implications The importance of correct diagnosis cannot be overemphasized as otherwise prolonged therapy and other unnecessary procedures may be instituted

Traumatic Periostitis in Young Children Traumatic periostitis in young children may ensue after a trivial traumatism and is relatively common It is associated with a limp and most often affects the tibia and the femur There is pain on palpation and limping but no swelling The first roentgen study immediately after the injury is negative but re-

distance into the cortex on one side to a complete fracture No displacement occurs Periosteal new bone formation is demonstrable in many instances at the time of the initial examination although it may be so faint that it can be demonstrated only by technically perfect films In some cases the fracture line can be demonstrated only with the extremity in partial obliquity In many instances there is a laminated appearance of the periosteal new bone In the early stages a small localized area of fraying-out of the margin of the cortex may be the only manifestation Subsequent examinations reveal the fracture line and callus The fracture may be bilateral Callus may be present along the lateral aspect of the fibula at the level of the fracture of the tibia, although no fracture line may be demonstrable in the fibula The lesion is easily confused with an infectious or neoplastic process Therapy consists of immobilization in a plaster cast for about six weeks followed by physiotherapy and graduated exercise In the case of a soldier, he usually cannot return to duty for two to three months In some instances there may be persistent pain and tenderness which require mobilization for longer periods

Fatigue or March Fracture of the Fibula Fatigue fractures of the fibula are apt to occur in young male skaters runners soldiers and active or hard working women of middle age and over It usually involves the slender cancellous bone slightly above the external malleolus In the upper portion of the fibula the fracture is most often brought on by jumping In some instances the fracture line is evident within a week although in others it is not demonstrable until the second or third week A band or rarefaction is visible at the end of twelve to eighteen weeks In children callus develops as early as the eighth day Adults show callus during the third week At twelve to sixteen weeks it forms a spindle like thickening crossed by a less dense stripe at the site of the fracture A dense band is often the first indication of the fracture and may be the last manifestation to disappear The plane of the fracture is almost horizontal

THE DELAYED APPEARANCE OF FRACTURES

Fractures may not be demonstrable roentgenographically until several days or weeks after a trauma This is particularly true of the flat bones such as the ribs transverse processes sternum navicular facial bones and vertebræ It may be due to the fact that the fracture line is so fine or lies at such an angle that it is not demonstrable even in well executed roentgenograms After a lapse of time there is absorption of bone or slight displacement of the fragments which makes the fracture clearly visible on the roentgenogram In every case with clinical evidence of fracture a negative examination particularly if the study has been performed soon after the trauma must not be accepted as final and the roentgen examinations should be repeated one or more times at intervals of a few days or weeks It seems improbable that a patient can sustain a fracture without knowing that there had been an injury sufficient to warrant medical treatment Such cases do occur however and may involve the bones of practically any part of the body In medico-legal cases the attorney frequently stresses the fact that a patient could not have had a fracture and ignored or forgotten it Nevertheless if roentgen examination shows evidence of an old fracture such evidence should be accepted in spite of the patient s having forgotten the injury Often the old fractures are

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examination after seven to ten days shows evidence of an ossifying periostitis This may persist for many weeks, disappearing usually in three to four months The lesion is also seen in the proximal femur as a result of birth trauma It is frequently mistaken for primary neoplasm, a serious error in diagnosis

Kummel's Disease Delayed Post-traumatic Spondylitis In 1891 Kummell described a clinical entity in which a trivial trauma to the spine was followed by an indefinite asymptomatic period and eventually culminated in symptomatic, progressive angular kyphosis The sequence of injury, latent period, and symptomatic progressive angular kyphosis is known as Kummell's disease The symptom complex may occur at any age though it is rare in young people Kummell's disease or post-traumatic

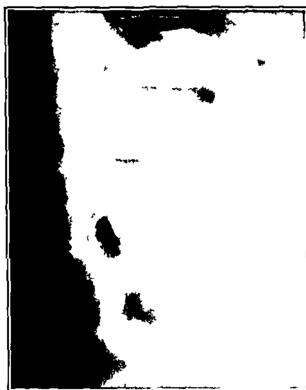


FIG 145 Old Compression Fractures of the Spine Kummell s Disease The bodies of the first and second lumbar vertebræ show anterior wedging with irregularity of the anterior and superior surfaces The patient was a male fifty six years of age with a history of a fall from a tree forty years ago

spondylitis comprises delayed collapse of the affected vertebræ and is believed to be due to an injury of the osseous and ligamentous structures of the spine which causes interruptions of the bony continuity and interference with the blood supply The diagnosis can be established definitely only by negative roentgenograms immediately after the trauma and later roentgen study showing positive evidence of anterior vertebral wedging and collapse Trauma to the spine however trivial, constitutes a definite indication for roentgen study A marked kyphosis is usually present and there is also spinal osteoporosis of varying degrees There is pain and disability with in rare instances reflex changes and paralysis The narrow portion of the vertebral wedge is anteriorly the posterior aspect usually being preserved The intervertebral spaces are normal in width or slightly narrowed

Differential diagnosis must include tuberculosis, herniated nucleus pulposus, back strain, ankylosing spondylitis, and hypertrophic arthritis. Therapy comprises stabilization of the affected vertebra to prevent increasing kyphosis. The prognosis is good.

OSTEOPOROSIS

Osteoporosis is defined as an absorption of bone with the result that the tissue becomes unusually porous and fragile and is characterized by a decrease in bone mass due to a lessened production of osteoid by the osteoblasts. The roentgen and pathologic concepts of osteoporosis differ markedly. To the roentgenologist, osteoporosis is an indication of a deficiency of calcium in the bone and is manifested by rarefaction of the bone. The condition is due to destruction of bone by the osteoclasts with resultant thinning of the trabeculae, the Haversian systems of the cortex undergoing enlargement. In rickets and osteomalacia there is actually a decrease in the calcium content of the affected bone. The bone contains excessively large amounts of non-calcified osteoid tissue and analysis of the bone reveals an abnormally low calcium content. Whether the calcium has been actually absorbed from the bone or the bone has never become fully calcified cannot always be determined on the basis of the roentgen manifestations alone. When the calcium has been absorbed or "abstracted" it is termed decalcification, while failure of calcification to ensue constitutes hypocalcification. In normal bone, the osteoblasts are continually building a protein matrix within which a calcium phosphorus-carbonate salt is deposited. In true osteoporosis, there is a decrease in the bone density due to interference with the process of matrix formation. The process may occur as the result of: (1) lack of stress and strain, (2) protein deficiency, and (3) steroid imbalance. It occurs in the post-menopausal period, during senility, and in diseases such as diabetes mellitus, Cushing's syndrome, and hyperthyroidism. It may also develop without known cause in young persons and in these instances is termed idiopathic osteoporosis. The osteoporosis of old age is probably due to the decrease or absence of gonadal hormones and the adrenocortical N hormone and shows a response to estrogen and testosterone therapy. In the post-menopausal state it is attributable to lessened estrogen production and also responds favorably to estrogen and testosterone. The syndrome is caused by failure of gonadal secretion of steroids which physiologically produce stimulation of osteogenesis. It is known that the estrogens or androgens have a favorable effect on the retention of calcium. In the adult, months or years may be required for the replacement of calcium deficiency. This accounts for the evidence of clinical improvement without demonstrable change in the appearance of the bones as shown by the roentgenogram. Osteoporosis is a disease of tissue metabolism and not of calcium metabolism. In consequence it is associated with relatively normal serum calcium and phosphorus levels. Calcium and Vitamin D lack in themselves probably do not cause osteoporosis.

Osteoporosis, decalcification, or loss of bone substance is a very important manifestation as it may be the only alteration which gives a clue to the diagnosis. In many instances the presence of osteoporosis represents the earliest or first deviation from the normal and enables the radiologist to establish the diagnosis of a disease process before more

serious and irreversible changes have taken place Thus, in early tuberculosis of the joint decalcification may be demonstrable in the bones about the affected joint prior to the development of actual destructive changes in the articulation The establishment of the diagnosis and the institution of proper therapeutic measures at this stage of the disease may prevent the progress of the lesion and result in cure or marked lessening of the damage to the affected structures In similar fashion, the presence of osteoporosis may supply the essential data to permit of a diagnosis of an obscure disease such as osteomalacia, hyperparathyroidism, and similar glandular and metabolic disturbances

Ostoeporosis may be limited to a solitary bone or portion of a bone, may affect several bones, or involve the entire skeleton The degree, extent, and character of the change are of great importance in determining the cause and the establishment of the diagnosis The most common cause is disuse atrophy the changes affecting the bone or bones in the region of and distal to the site of the lesion The osteoporosis is reversible, usually disappearing promptly and completely after the re establishment of normal use of the affected portion of the body Endocrine disturbances as in hyperparathyroidism afford a classical example of osteoporosis There is increased absorption and simultaneously decreased deposition of calcium The changes are associated with localized cystic decalcifications and absence of the bone trabeculations In diabetes mellitus, there is generalized decalcification because of the increased resorption of calcium from the bones This is associated with prolonged acidosis which results in depletion of the alkali reserve of the body Hyperthyroidism is associated with diffuse osteoporosis and is characterized by generalized cortical thinning, diminution of the cancellous structure, and increase in the width of the medulla Disorders such as osteogenesis imperfecta and osteomalacia result in a decrease or lack of deposition of minerals with generalized and severe decalcification Leprosy and other diseases with involvement of the nerves supplying the bones termed neurotrophic disorders are associated with severe degrees of osteoporosis The affected bones in addition to the osteoporosis may in some instances show multiple small cyst-like areas of radiance in the affected zones Vascular spasm such as occurs in Raynaud s disease and similar conditions results in symmetrical, progressive decalcification The terminal phalanges may be absorbed partially or completely Acute and chronic osteomyelitis is associated with cortical thinning due to a decrease in the bony lamellæ and an actual resultant increase in the width of the medullary cavity Osteitis deformans presents areas of osteoporosis This is well exemplified by the process termed osteoporosis circumscripta which occurs particularly in Paget s disease of the skull and other flat bones Periosteal neoplasms and primary or secondary bone tumors of other types are associated frequently with irregular localized or diffuse osteoporosis With increased growth of the tumor in the medullary spaces in the Haversian canals bone lamellæ undergo compression separation and destruction with replacement by tumor tissue Pressure from extra osseous tumors may produce varying degrees of osteoporosis also Many other causes of osteoporosis can be enumerated Loss of blood supply results in aseptic necrosis Aneurysms may cause pressure destruction of adjacent bone In tuberculosis the reticuloses bone cysts hyperplasia of the bone marrow in blood dyscrasias gouty tophi fibrocystic disease of the pancreas and similar conditions, bone tissue is replaced by fibrous tissue or otherwise

altered with resultant osteoporosis Osteoporosis may occur in association with extensive destruction of the parenchyma of the liver and results from toxic influences occlusion of the bile ducts, or external biliary fistula This is termed hepatogenic osteoporosis In cirrhosis, jaundice may be accompanied by osteoporosis Malignant liver disease, either primary or metastatic with occlusion of the bile ducts leads to osteoporosis The mechanism of the osteoporosis is varied and complex In nearly all cases

Fig 146 Fig 147





Fig 148

Fig 146 Marked Osteoporosis There is marked cortical thinning increased radiance and practically complete absence of the bony trabeculæ The osteoporosis developed subsequently to a fracture dislocation of the upper end of the humerus in a female seventy years of age

Fig 147 Compression Fracture of the Ninth Dorsal Vertebra (arrow) Marked osteoporosis of the spine

Fig 148 Advanced Osteoporosis in Fibrocystic Disease of the Pancreas There is an advanced degree of osteoporosis of all the bones manifested by cortical thinning increased radiance and diminution of the trabecular pattern The soft tissues show marked wasting Study of the chest showed emphysema and peribronchial thickening involving both lungs

there are disturbances of calcium and fat resorption from the intestines and faulty absorption and deposition of vitamin D in the liver Associated secondary manifestations comprise hyperparathyroidism due to lack of calcium, atrophy of the testes due to failure of inactivation of estrogens by the liver, and hyperplasia of the adrenal cortex Histopathologic examinations reveal simple osteoclastic osteoporosis or bone resorption with and without osteoid bands The roentgen findings are those of diffuse osteoporosity chiefly of the vertebral and pelvic bones and less commonly of the long bones and ribs The dorsal vertebræ tend to be wedged and the lumbar vertebræ become fish shaped

The clinical manifestations of osteoporosis vary widely The initial complaints are usually weakness and pain in the back and hip Roentgen examination reveals compression of one or more of the vertebral bodies In the dorsal region there is usually a wedge shaped deformity while in the lumbar region there is a tendency to general narrowing The density of the vertebral bodies in the advanced cases becomes so slight that only a faint outline may be visible The opposing surfaces become concave, particularly in the lumbar region The porosis is less marked in the cervical region, the pelvis and the upper femurs The extremities are affected only in advanced cases The skull and the lamina dura remain normal in appearance A moderate degree of osteoporosis of the spine appears to be physiologic after the menopause and in most instances does not produce pain Pain when present results from secondary collapse of the vertebræ or ballooning of the intervertebral discs together with invagination of the vertebral plates Elderly persons as a rule also have osteoarthritis of the spine which may cause pain, although the pain in osteoarthritis is not as severe as that accompanying actual collapse or crushing of an osteoporotic vertebra Patients with postmenopausal osteoporosis frequently suffer fractures which are slow to heal Administration of estrogenic substances usually results in rapid improvement with healing of the fractures and recalcification of the bones Cessation of the estrogen treatment causes an exacerbation of the symptoms and resumption of the treatment is again followed by marked improvement

Post-traumatic Painful Osteoporosis Sudeck's Atrophy

Sudeck in 1900 was the first to describe the trophic changes in the bones of the hand and feet which may ensue after trauma His original report was concerned with acute atrophy of bone in association with inflammatory processes in the articulations He later described a type of osteoporosis which he called post traumatic reflex atrophy of bone and clearly differentiated this phenomenon from the bone atrophy described by Virchow and others as a result of tabes dorsalis syringomyelia and other central nervous system diseases Inactivity alone cannot account for the severe degree of bone changes particularly as the atrophy develops rapidly and while the extremity is in use Bones of the extremity which have been at rest for a period of time lose much of their mineral content and become more permeable to the roentgen rays This type of bone atrophy is of relatively little clinical significance as the bones rapidly regain their normal density and strength after normal function of the extremity is resumed During the period when the atrophy of the bone is marked the patient has no symptoms referable to this change In contrast to this form of atrophy post traumatic painful osteoporosis is a



FIG 149 Post traumatic Painful Osteoporosis Sudeck s Atrophy There is marked osteoporosis of the bones of the right foot The changes were consequent upon a fracture of the lower end of the fibula The changes are better appreciated by comparison with the normal left foot



FIG 150 Sudeck s Atrophy of the Hand and Wrist Post traumatic painful osteoporosis

major cause of prolonged loss of function or severe pain after injury to an extremity The patients are frequently accused of malingering because the onset of the disability is long after the effects of the original trauma should have disappeared or in the case of a fracture the bones have had sufficient time to become definitely united The early recognition of the roentgen changes is of the utmost importance if the period of disability is to be lessened A long period of diasbility results in economic loss and great psychic damage Unless the changes in the bones are arrested or cured, permanent damage develops

Clinical Manifestations Post traumatic osteoporosis is characterized by partial loss of function, pain, and vasomotor and trophic changes in the affected extremity The disturbance of function and pain are disproportionate to the injury The pain is not relieved by immobilization as occurs with fracture and infection The trauma which precipitates post traumatic osteoporosis may vary markedly in severity and type In some instances, the condition develops after a slight or moderate trauma to a polyarticular region such as the wrist or ankle The osteoporosis develops gradually during a period of three to four weeks and is associated with edema of the extremity The limitation of motion in the adjacent joints increases rapidly The differentiation between acute osteoporosis and arthritis is difficult The decalcification of the small bones of the affected extremity without other evidence of arthritis establishes the diagnosis although the two conditions may appear very similar Sudeck's atrophy may develop after a fracture of one or more of the bones of an extremity Any patient in whom pain does not disappear after reduction of the fracture and immobilization should be studied by roentgen methods at intervals of about two weeks to determine possible development of true osteoporosis It is important to make comparative studies of the normal limb in order to permit of accurate comparison of the structure of the bones particularly in the early stages Post-traumatic painful osteoporosis develops most often in the short bones of the hands and feet The epiphyses of the metatarsals the metacarpals, the phalanges and the long bones are frequently affected The diaphyses of the long bones rarely are involved The flat bones of the skull may be the seat of similar rarefaction and Schiller in 1929 described post traumatic osteoporosis of the calvaria There are no accurate figures as to the frequency of the condition Some authors have stated that it occurs in 50 per cent of those with trauma to the small bones of the hand and foot This is much higher than is usually believed to be the case

Roentgen Manifestations There are two principal forms of Sudeck's osteoporosis the acute and the chronic types The acute form is characterized by a mottled appearance of the bone due to irregular rarefaction of the spongiosa The mottling is usually most marked in the carpal and tarsal bones and in the heads of the metatarsals and metacarpals In advanced cases the cortex of the small bones is markedly thinned the outline of the individual bones becoming so indistinct that clear differentiation is impossible The lamella fade into each other and produce an ill defined homogeneous shadow In the chronic form the trabecula of the affected bones become very fine and difficult of recognition The margins of the bones are visible despite a generalized loss of mineral salts In the short bones, particularly the carpals and tarsals the process presents several stages of evolution At the onset the bones present diffuse mottling and increased radiance The areas of rarefaction become more extensive and the bones become uniformly permeable to

the arm Diffuse and marked demineralization indicates the height of the disease There is thinning of the cortex of the bones and longitudinal streaks make their appearance in the thin cortex In the carpal and tarsal bones, the cortical thinning results in the disappearance of the osseous margins and transforms the entire bone into a homogeneous mass At this stage, a diagnosis of tuberculosis or osteoarthritis is frequently made As the process undergoes regression, the margins of the small bones again become visible, the longitudinal lamellae become thickened, and there is gradual reappearance of calcium in the bone The reconstructive process is slow In some cases recalcification never takes place Complete restoration of the density of the bone is not necessary for symptomatic relief

Treatment Post-traumatic painful osteoporosis in some instances is a self limited disease and after a few weeks or months recalcification takes place spontaneously After the disease has reached the stage of practically complete decalcification improvement may begin spontaneously However even after the lapse of months or years, thinning of the cortex and lamellae with irregular areas of decalcification may persist The vasomotor manifestations and pain may disappear without therapy In these cases the recovery of function of the affected limb requires many months and during the stage of recalcification extensive fusion of the carpal or tarsal bones may ensue Ankylosis is a serious complication Since the milder forms of the disease may heal spontaneously and cause no permanent disturbance of function, it must not be assumed that a particular form of therapy comprises the true remedy for the disease in all circumstances Most cases do not undergo regression and must be treated to prevent permanent damage and disability Treatment in the past was symptomatic Sudeck recommended active motion Heat, massage, and voluntary movement are helpful Forceful manipulation under anesthesia is contraindicated Fixation or limitation of motion by orthopedic appliances aggravates the pain Roentgen therapy results in symptomatic relief but the absorption of bone and disturbance of function are unaffected and the period of disability is not shortened Intensive physiotherapy and functional stimulation are unsatisfactory The most effective form of treatment, particularly in the acute stage, is denudation of the main artery to the affected extremity, so called periarterial sympathectomy The nerve fibers in the adventitia of the large arteries of the extremities have a definite trophic function Interruption of abnormal trophic reflexes which course along these nerve fibers accounts for the strikingly beneficial results subsequent to sympathectomy of the periarterial type The neurotrophic fibers in the adventitia of the larger arteries are distributed segmentally to the periarterial nervous network of the peripheral somatic nerves and do not pass through the regional sympathetic ganglia or chains This accounts for the failure of improvement after surgical removal of the regional sympathetic ganglia or interruption of the rami to the ganglia (ramisection) while marked relief results after denudation of the major artery to the affected part Periarterial sympathectomy affords relief of pain and edema in twenty four hours The impaired function improves within a few days and the patients are able to resume work after about three months This is in striking contrast to the group treated by other methods which requires nine months or longer for recovery Early therapy is important as many of the untreated cases develop ankylosis of one or more joints with resultant permanent disability The severity of the trophic changes in

the bones is not proportional to the intensity of the trauma Pain persisting after trauma to an extremity should lead to the suspicion of post traumatic osteoporosis and is an indication of roentgen study of the affected extremity at frequent intervals in order to permit of early diagnosis The disease is frequently overlooked

Osteoporosis of Immobilization

Prolonged recumbency produces widespread skeletal alterations, particularly in conditions which require immobilization in the recumbent position for long periods of time and consequently exert an injurious influence upon calcium and nitrogen metabolism The incidence of urinary lithiasis is increased due to increase in excretion of calcium, sulphur, nitrogen, and phosphorus with resultant negative nitrogen balance The pale wizened, hunch back child after prolonged immobilization for tuberculosis of the spine is familiar The patients are stunted to a greater degree than should occur from the kyphosis alone Loss of calcium beyond a certain degree and chronic disturbance of nitrogen metabolism may produce permanent changes in the tissues Stevenson studied the widespread effects of immobilization upon the skeleton the effect of immobilization and recumbency in patients with chronic disease in whom porosis previously had not been present and the changes in the lower limbs after immobilization He attempted to determine the common factor in these effects Prolonged immobilization even though the limbs are entirely free from disease produces gross roentgen changes in the lower limbs due to disturbance of calcium metabolism Immobilization of one lower limb produces osteoporosis of the entire hind quarter The changes in children and young adults may be permanent Ambulatory patients may have subacute tuberculous arthritis for months or years without developing osteoporosis On immobilization a marked degree of osteoporosis develops Similar patterns are present throughout the leg proximal to a diseased tarsus distal to a pathological hip or in the opposite limb which has been immobilized Bilateral changes occur in the legs in the presence of a lesion of the spine The changes are the same following immobilization for tuberculous arthritis Still s disease rheumatoid arthritis or traumatic synovitis and anterior poliomyelitis

Roentgen Manifestations There is generalized decrease of bone density a zone of more intense porosis at the original site of the metaphysis a narrow zone adjacent to the cortex of the bone ends and irregularly distributed patches Bone or cartilage destruction permits of differentiating by roentgen methods of the changes in immobilization osteoporosis in a diseased joint and an otherwise healthy joint Intense local osteoporosis may cause practically complete roentgen disappearance of bone The porotic bone may recalcify or may disintegrate and be lost irrevocably In joints which on roentgen examination appear most severely affected the actual tuberculous process may be limited to the surface of the bone or extend only for a distance of two or three millimeters Therefore in some cases the loss of cancellous bone may be due to mechanical disintegration during the stage of intense osteoporosis rather than to destruction by tuberculous granulation tissue Recalcification at the site of disease while the part is immobilized is an important manifestation of healing A positive calcium balance hastens healing by facilitating recalcification and is important to prevent the mechanical collapse of osteoporotic cancellous bone

JOINT PROSTHESIS

In 1948 the Judets introduced the new technique in which the femoral head is excised and replaced by an acrylic head which is fixed firmly to the remaining part of the neck of the femur by a long stem In the Judet technique the artificial head moves only in the acetabulum While the results seem better with this method, the cases have not yet been observed for a sufficient length of time to permit of a definite conclusion Valls suggests a modification in which the stem is fluted or flanged in the form of a three flanged nail The method suggested by Valls utilizes the same shape as the normal head of the femur, but unlike the acrylic head it is hollow and the flanged stem destroys only a very small amount of bone of



Fig 151 Metallic Prosthesis *A* Anteroposterior view *B* Lateral view The head and neck of the femur have been replaced by a metallic prosthesis The prosthesis is maintained in position by an intramedullary pin

the neck The method of Smith-Petersen, based on the use of a vitallium mold has the advantages of lessening the absorption and wearing away of the neck of the femur and the anatomy of the joint is much more normal than with the previous operations The basis of the Judet operation is resection reconstruction and consists of excision of the pathological head of the femur and its replacement by an artificial head made up of a synthetic plastic material which is firmly fixed in the upper end of the femur The prosthesis is made of an artificial substance which is well tolerated by the tissues of the body and does not produce an unfavorable reaction in the surrounding tissues The acrylic head of Judet requires an extensive resection of the bone The cylindrical stem destroys approximately a third of the diameter of the head of the femur and in some instances very little bone remains Some orthopedic surgeons use a resin like substance which is nonopaque to the x ray This has the advantage of allowing complete examination of the structure of the bone in contact with the prosthesis In the case of the hip the operation is indicated for osteo-

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arthritis ununited fractures of the femoral neck nontuberculous ankylosis and dislocation of the hip and is applicable to patients of any age It has been used in various joints of the body and is now well established as a procedure in orthopedics

Prostheses for the femoral head and other parts of the body have come into widespread use but have not been utilized long enough to justify final evaluation of their usefulness in the treatment of painful pathological disabilities of the joints The use of certain of these devices has in many instances resulted in early good function and great comfort The hip joint will tolerate weight bearing and motion and thrive when forces are brought to bear upon it through a foreign body such as a vitallium cup which moves freely between the acetabulum and the head of the femur Great interest has been shown in prosthetic devices by orthopedic surgeons and the radiologist must be familiar with the various types used both those which are opaque and non-opaque to the x ray The use of prostheses for the head of the femur is still in the stage of discussion These methods represent a new approach to fusions osteotomies and other attempts to meet the problem of the painful unstable or fractured hip

Despite a widespread enthusiasm for the procedure there are certain drawbacks which are beginning to be appreciated Laing described the pathological changes in the neck of the femur in a patient who died two weeks after the performance of the operation The prosthesis was firmly imbedded in the femoral neck and roentgen study of the specimen showed no rarefaction of bone around its stem A block of bone from the postero-medial aspect of the femoral neck was removed close to the overlap of the prosthesis the internal margin of this block comprising the bone in intimate relation to the stem of the prosthesis and roentgen study was performed by crystallography tube The radiographs revealed three main zones consisting of outer and inner trabeculated areas with dense shadow intervening In the first or inner zone the cancellous bone presented a corrugated surface at the point of contact with the stem of the prosthesis The middle zone consisted of the dense compact bone of the calcar femorale the vertical lamina of strong tissue in front of the lesser trochanter serving to strengthen the neck of the femur The outer zone was composed of external trabeculated bone The histological changes varied in each of these zones The appearances were those of extensive bone necrosis involving the calcar femorale an important weight-bearing area of the femoral neck The necrosis was not of the block infarct type being more irregular in its occurrence and had not elicited a marked inflammatory reaction The changes indicated extensive necrosis surrounding the stem of the prosthesis and emphasized certain advantages and disadvantages of the use of prostheses

The use of prostheses has a small mortality in careful hands averaging less than 2 per cent Hematoma formation may occur Wound infections develop in some of the patients and necessitate removal of the prosthesis The presence of infection indicates that the results will be poor as regards function A serious complication is persistent pain This may be due to faulty insertion of the prosthesis Contraindications to the operation comprise senility obesity circulatory changes in the leg and certain general disease Faulty position of the stem with relation to the neck of the femur produces pain and coxa valga An excessively long stem with projections from the subtrochanteric cortex may cause pain Painful sclerosis of the bone may be caused by pressure of the stem along the medullary surface of the cortex Thrombophlebitis post-operative

dislocation, fracture of the stem of the prosthesis and undue wear on the surface of the head also cause poor results

ADDITIONAL READING

JUDET R and JUDET J Technique and Results with the Acrylic Femoral Head Prosthesis J Bone & Jt Surg 34B 173 1952



Fig 152 Intramedullary Prosthesis There is a long metallic intramedullary prosthesis in the lower fibula A metallic screw and wire are visualized in the lower end of the tibia



Fig 153 Prostheses of Acrylic Type The patient had advanced rheumatoid arthritis with extensive destruction of both hip joints Prostheses were utilized to replace the femoral heads The prostheses are of the acrylic type and are non opaque to the x ray only the metallic nails being visualized on the roentgenogram

BONE GRAFTS

The replacement of absent segments of bone by implantation of bone or a substitute for bone is termed bone grafting. The results of the procedure conform with our knowledge of bone repair and functional adaptation and reconstruction of bone. There are four methods of repair of defects of bone. (1) The method is termed autoplastic if the bone of the person or the experimental animal is transplanted in the defect. (2) When the bone of the same species is grafted from one individual to another the grafting is termed homoplastic. (3) The graft is called heteroplastic if bone of a different species is used. (4) Alloplastic is the term applied in cases in which instead of bone a substitute such as ivory or a mixture of calcium salt is employed. The most favorable results are obtained with autoplastic grafts. The vitality of transplanted bone is dependent on the surface area of the transplant which remains in contact with living tissue. A transplant covered by periosteal and endosteal connective tissue, particularly when comprised in part of spongy bone, has the best chance of survival as the circulation of blood in the connective tissue overlying the transplant is speedily restored and the bone receives nutrient material sooner. Even under the most favorable conditions a large portion of the implanted bone tissue may undergo necrosis. This does not necessarily retard the healing of the defect. The implanted bone is gradually replaced by new bone since the structure of the implanted bone is never entirely adequate as regards function.

DISLOCATIONS

Dislocations are most commonly encountered in young adults and the middle decades of life. During childhood the epiphyses are unfused and the bones are relatively soft, hence a trauma is most apt to result in epiphyseal separation or fracture. Elderly people suffering an injury usually develop a fracture as the bones are relatively weak. In adult life the bones are strong and the capsule is the weakest link, hence trauma during this period is apt to cause a dislocation. Dislocations are not infrequently reduced prior to roentgen examination as the deformity and disability are so typical that clinical diagnosis is definite. This is not a safe practice. Every dislocation should be examined roentgenographically before treatment is instituted. Fractures may complicate the dislocation with resultant severe damage to the soft tissues during attempted reduction. The surgeon may be blamed for having caused the fracture during reduction with resultant liability to suit for damages. Severe fractures in the region of a joint with marked displacement of fragments may closely simulate a dislocation and result in an erroneous diagnosis. A slipped epiphysis may easily be mistaken for a dislocation without roentgen study. Chronic or recurrent dislocations may require special studies in an attempt to determine the underlying cause of the condition. Secondary dislocations may occur in association with Charcot's disease, tuberculosis and other lesions in or about the joint. In addition to the usual projections, oblique, stereoscopic and other views may be necessary. Comparison of contralateral joints is of value in doubtful cases.

Dislocation of the Shoulder. Dislocation of the shoulder is the most common of all dislocations. The dislocation may be either subglen-

oid or subcoracoid In the typical case the head of the humerus is displaced downward, the glenoid is visible with no overlying humeral head, and the distance from the acromion process to the humerus is increased Proper positioning of the patient during the roentgen examination is essential to prevent error in diagnosis Roentgen studies with the arm flexed across the chest or in other abnormal position may result in incorrect diagnosis Fractures of the neck or tuberosity of the humerus occur frequently in association with dislocations of the shoulder It is essential that regardless of the obviousness of the diagnosis clinically, roentgen studies be carried out before reduction is attempted Injuries to the soft tissues frequently result in extensive calcific depositions about the shoulder joint Posterior dislocation of the shoulder is rare and is frequently overlooked



Fig 154 Dislocation of the Shoulder There is complete dislocation of the shoulder the head of the humerus lying below the glenoid

or diagnosed incorrectly The clinical and roentgen manifestations of the condition are characteristic although they must be carefully sought for in many cases There is decreased palpable anterior prominence of the head of the humerus, increased prominence of the coracoid and the humeral head below the acromion limitation of abduction and absence of external rotation In most instances there is a considerable delay before the diagnosis is established This is probably explained by the rarity of the injury which results in its being unsuspected and also by the fact that the condition is not recognized because the physical signs are masked by swelling A single anteroposterior view of the shoulder is entirely inadequate Stereoscopic roentgenograms may be helpful Transthoracic lateral or oblique projections with the patient erect establish the diagnosis with definiteness

Dislocations of the Acromio-clavicular Joint While considered by many surgeons to be relatively unimportant, this lesion may be the cause of much pain and disability It occurs most commonly in laborers as the result of a severe pull of the arm, in lifting heavy objects, and in sports, especially football Routine films of the shoulder may demonstrate an increase in the width of the acromio clavicular space or upward displacement of the lateral end of the clavicle With the patient lying down, the joint may appear identical with its fellow on the opposite side By carrying out the examination with the patient in the erect position and

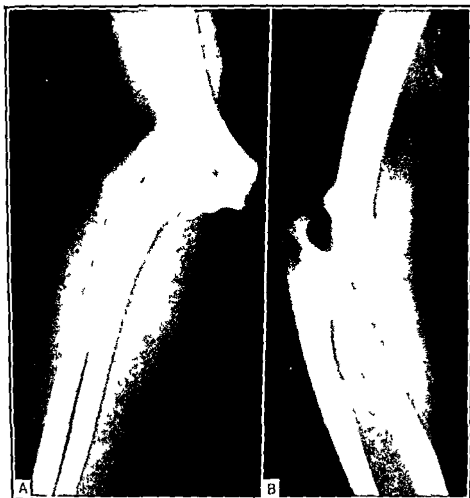


FIG 153 Dislocation of the Elbow *A* Anteroposterior view *B* Lateral view The humerus is displaced anteriorly and medialward No fracture of the bones is demonstrable

holding a weight in the hand the dislocation becomes clearly demonstrable and this position must be used in all suspected cases Both sides should be examined for comparison

Dislocation of the Elbow Dislocation of the ulna is the most common dislocation about the elbow The lower end of the humerus lies in contact with the anterior aspect of the upper end of the ulna Fractures of the coronoid process of the ulna may be associated with the dislocation Dislocation of the radius occurs in association with dislocation of the ulna or as a solitary lesion Fracture of the ulna in the region of the upper third of the shaft not infrequently occurs simultaneously

Dislocation of the Radio-ulnar Articulation The ulna may be displaced laterally or dorsalward particularly in association with fractures and other severe injuries of the wrist. In Madelung's deformity, there is dislocation of the ulna due to interference with the growth of the lower epiphysis of the radius.

Dislocation of the Lunate The commonest dislocation at the wrist joint involves the lunate bone. This is caused by falls with the hand bent backward and hyperextended. The trauma is similar to that which produces a Colles' fracture and it may occur in association with this condition or fracture of the navicular. The displaced bone can usually be reduced relatively easily if reduction is performed soon after the trauma-



FIG 156 Dislocation of the Lunate There is complete dislocation of the left lunate the articuarl surface being directed anteriorly (arrow) The normal wrist is shown in similar projection

tism However the ligaments and tendons undergo shortening and after the lapse of a relatively short time operative reduction or removal of the dislocated bone becomes necessary. The roentgen diagnosis is established on the basis of two deviations from the normal. The lateral view reveals that the distal surface of the lunate, which normally articulates with the proximal aspect of the capitate faces directly toward the palmar aspect of the wrist. In the posteroanterior view, a space is present between the navicular and the triangular. Dislocations of the lunate are of two types perilunar and rotational. In the former the lunate remains in relatively normal position, the distal carpals being displaced backward and upward. In the rotational type the dislocation is usually toward the palm and is associated with tearing of the ligaments. If the anterior ligament is intact the lunate rotates forward and upward from 90 to

250 degrees Roentgen examination is essential to establish the diagnosis On clinical examination, the dislocated bone may be palpable, the palmar aspect of the wrist is altered and the fingers are held in slight flexion Immediate manipulation is the treatment of choice Immobilization for several weeks is essential Roentgen examination should be carried out at intervals of one to four months for a period of one year or longer to rule out aseptic necrosis

Dislocation of the Pisiform Dislocation of the pisiform is rare It may result either from a direct injury or muscular violence On clinical examination there is a depression at the site normally occupied by the pisiform Movements are painful and limited Demonstration of the lesion necessitates roentgenograms with the ulnar side of the wrist placed against the film in about 15 degrees of supinated rotation The bone may return to its normal position spontaneously Cases which do not respond to treatment require surgical removal of the fragment

Dislocations of the Hand Dislocations of the phalanges are common and may result from falls wrenches or trauma incidental to football basket ball, wrestling and similar sports The distal fragment may be displaced anteriorly laterally or posteriorly Chip fractures are not uncommonly associated with the dislocation Traction usually reduces the dislocation and not infrequently an athletic trainer or coach "snaps" the joint back into place immediately after the trauma However a severe tear of the capsule not infrequently occurs This requires open reduction Metacarpo phalangeal dislocations do not differ from similar lesions of the phalanges

Dislocation of the Hip Joint Dislocations of the hip are relatively rare and usually occur in adults between the ages of twenty and forty five years In the young, severe injuries to the hip region result in a separation of the epiphysis while in elderly people fracture of the neck of the femur usually ensues The dislocation is most commonly upward with the head of the femur lying at or above the level of the superior margin of the acetabulum Fracture of the acetabulum may accompany the dislocation Rarely, the femoral head is displaced downward in which case it overlies the obturator foramen In the presence of upward or downward dislocation of the head of the femur diagnosis is not difficult When the displacement is directly backward the relations of the acetabulum and head of the femur appear normal However comparison of the two sides shows that the head of the femur on the affected side is distinctly smaller and more sharply defined as it lies in closer relationship to the roentgen film than in the normal hip

Congenital Dislocation of the Hip Classical congenital dislocation of the hip is considered secondary to a genetic dysplasia of the acetabulum and develops during intra uterine life or the first or second post natal year There is aplasia or hypoplasia of the roof or buttress of the rim of the acetabulum which results in a flat shallow socket Dislocation of the femoral head ensues because the hypoplastic and insufficient roof of the acetabulum lies in the axis of transference of forces of body weight and muscle action Actual dislocation need not necessarily occur as dysplasia may constitute a permanent deformity with characteristic clinical symptoms and roentgen manifestations despite the fact that it is merely a precursor of the classical dislocation In a dysplastic and inadequate hip formed during fetal life various possibilities for the subsequent development of the joint must be considered In some

instances, the hip joint spontaneously returns to normal during intra uterine life or early infancy without treatment and the dysplastic hip with a shallow socket may persist throughout life with no dislocation of the femoral head ever taking place In other instances, the femoral head is partially displaced from the acetabulum as a subluxation This may persist or the displacement of the femoral head may gradually increase from a partial to a complete dislocation In rare instances, extreme degrees of dysplasia with irreparable malformation of all the structures of the hip joint and marked shortening of the shaft of the femur develop Congenital subluxation is a precursor of actual dislocation During the first six months of life, dislocation is rare The clinical manifestations in this period are related to contracture of the adductor muscles of the hip



Fig 157 Dislocation of the Hip There is dislocation of the right hip the head of the femur overlying the obturator foramen There is also a fracture of the right pubis (arrow)

The changes may become permanent and irreversible by the age of two or three years In the establishment of the diagnosis the clinical manifestations may be more significant than the roentgen manifestations, particularly during infancy Careful correlation of the clinical and roentgen studies is essential for early diagnosis

There is relatively little knowledge of the sequence of events in congenital dislocation of the hip prior to birth as knowledge of the embryological development of the normal hip is not complete The hip is well formed by the tenth fetal week including the acetabulum femoral head and neck and capsule The normal embryological development of the hip tends to ensure a stable joint Cases with dislocation of the hip before birth emphasize the theory of primary dysplasia of the acetabulum The cartilage of the superior margin of the acetabulum and the glenoid is plastic in the fetus and infant In subluxation the pressure of the head of the femur

against the glenoid and the superior margin of the acetabulum depresses these structures and a groove or fossa forms imparting an obliquity to the roof of the acetabulum Unless the pressure is relieved, the obliquity increases with increasing ossification and at about the age of five years the deformity becomes permanent Continued pressure of the head against the superior margin and simultaneous filling in of the base of the acetabulum cause the acetabulum to become elongated and shallow Complete dislocation with the femoral head lying above the acetabulum results in a small acetabulum, the obliquity of the roof becoming relatively slight or absent If the femoral head dislocates completely in the early months of life, there is no pressure exerted on the brim of the acetabulum and the obliquity of the roof remains as at the time of dislocation The constant pathological finding in congenital dislocation of the hip is elongation or relaxation of the capsule The cause of the elongation of the capsule is not known It may be genetic, mechanical, or hormonal in origin Some observers believe it is due to nutritional deficiency Congenital dislocation of the hip is not primary but is secondary to delayed and inhibited ossification of the cartilaginous structures which enter into the formation of the hip joint The changes involve both the ilium and the femur The term congenital dysplasia of the hip is logical as the anomaly affects the entire innominate bone Complete displacement of the femoral head from the inadequate acetabulum is not common Partial displacement or subluxation is a frequent cause of serious hip joint disability during early and adult life as it is often unrecognized

Roentgen Manifestations The roentgen manifestations comprise inhibited, delayed and altered endochondral ossification or osteogenesis of the innominate bone and femur with displacement of the femoral head from the acetabulum There is tilting of the pelvis and adduction of the affected extremity There is delay in the ossification of the cartilaginous roof of the acetabulum and the bony acetabulum is flat and inadequate Delay in ossification of the ilium and delayed ossification or closure of the ischiopubic cartilaginous juncture are important roentgen features There is inhibited and altered osteogenesis of the cartilaginous femoral head The degree of dysplasia varies in the component anatomic elements of the hip joint The dysplasia of the acetabulum may persist while the dysplasia of the head may increase during the period of disappearance of the acetabular dysplasia Pelvic obliquity and adduction of the extremity are almost invariable roentgen manifestations and are secondary to the postural shortening or contracture of the adductor muscles, tendons and fascias During the early years of life the cardinal roentgen manifestation is an abnormal acclivity of the roof of the acetabulum with increase in the angle of incidence of the roof of the socket There is an increase in the distance between the upper femoral diaphysis and the acetabular floor with hypoplasia or delayed development of the epiphyseal nucleus of the femoral head Because of repeated trauma resulting from abnormal shearing forces mechanical instability and other concomitants of the dysplastic hip secondary changes develop These comprise abnormalities of development of the bone and soft tissues and arthritic sequelae When the dislocation is reduced early in life a well formed acetabulum may be attained as a result of the weight bearing thrust of the head of the femur

In the fully established case the acetabulum becomes elongated and shallow and *the femoral head lies outside the joint and assumes a flat* or conical shape The neck of the femur is broad and short and there is

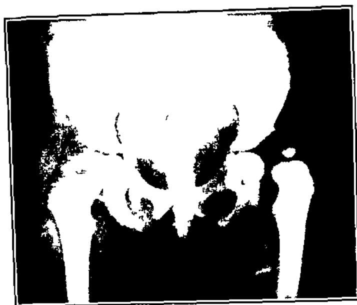


Fig 158 Congenital Dislocation of the Hip The left acetabulum is shallow The head of the left femur lies at a higher level and more laterally than normally The bones of the left side of the pelvis and the left upper femur show marked underdevelopment and are smaller than the corresponding portions on the right The soft tissues about the left hip are distorted due to the abnormal position of the left femur



Fig 159 Congenital Dislocation of Both Hips in an Adult Both hips are completely dislocated The true acetabulums are shallow The heads of the femurs are displaced markedly upward and are small and irregular in outline The femoral necks are short and form almost a right angle with the shafts The pelvis is tilted The bones of the pelvis and the femurs are decreased in size The patient a female forty years of age knew that the condition had existed since birth No attempt had ever been made to correct the dislocations

against the glenoid and the superior margin of the acetabulum depresses these structures and a groove or fossa forms imparting an obliquity to the roof of the acetabulum Unless the pressure is relieved, the obliquity increases with increasing ossification and at about the age of five years the deformity becomes permanent Continued pressure of the head against the superior margin and simultaneous filling in of the base of the acetabulum cause the acetabulum to become elongated and shallow Complete dislocation with the femoral head lying above the acetabulum results in a small acetabulum, the obliquity of the roof becoming relatively slight or absent If the femoral head dislocates completely in the early months of life, there is no pressure exerted on the brim of the acetabulum and the obliquity of the roof remains as at the time of dislocation The constant pathological finding in congenital dislocation of the hip is elongation or relaxation of the capsule The cause of the elongation of the capsule is not known It may be genetic mechanical, or hormonal in origin Some observers believe it is due to nutritional deficiency Congenital dislocation of the hip is not primary but is secondary to delayed and inhibited ossification of the cartilaginous structures which enter into the formation of the hip joint The changes involve both the ilium and the femur The term congenital dysplasia of the hip is logical as the anomaly affects the entire innominate bone Complete displacement of the femoral head from the inadequate acetabulum is not common Partial displacement or subluxation is a frequent cause of serious hip joint disability during early and adult life as it is often unrecognized

ROENTGEN MANIFESTATIONS The roentgen manifestations comprise inhibited delayed and altered endochondral ossification or osteogenesis of the innominate bone and femur with displacement of the femoral head from the acetabulum There is tilting of the pelvis and adduction of the affected extremity There is delay in the ossification of the cartilaginous roof of the acetabulum and the bony acetabulum is flat and inadequate Delay in ossification of the ilium and delayed ossification or closure of the ischiopubic cartilaginous juncture are important roentgen features There is inhibited and altered osteogenesis of the cartilaginous femoral head The degree of dysplasia varies in the component anatomic elements of the hip joint The dysplasia of the acetabulum may persist while the dysplasia of the head may increase during the period of disappearance of the acetabular dysplasia Pelvic obliquity and adduction of the extremity are almost invariable roentgen manifestations and are secondary to the postural shortening or contracture of the adductor muscles tendons and fascias During the early years of life the cardinal roentgen manifestation is an abnormal acclivity of the roof of the acetabulum with increase in the angle of incidence of the roof of the socket There is an increase in the distance between the upper femoral diaphysis and the acetabular floor with hypoplasia or delayed development of the epiphyseal nucleus of the femoral head Because of repeated trauma resulting from abnormal shearing forces mechanical instability and other concomitants of the dysplastic hip secondary changes develop These comprise abnormalities of development of the bone and soft tissues and arthritic sequelæ When the dislocation is reduced early in life a well formed acetabulum may be attained as a result of the weight bearing thrust of the head of the femur

In the fully established case the acetabulum becomes elongated and shallow and the femoral head lies outside the joint and assumes a flat or conical shape The neck of the femur is broad and short and there is

the femur and patella lie anteriorly and overlap the tibia Lateral projections best show the nature of the lesion and the position of the bones Fractures seldom occur in association with dislocations of the knee Dislocation of the patella may occur with lateral displacement of the patella The anteroposterior view is important to demonstrate the condition Dislocation of the semilunar cartilage is not demonstrable as the cartilage is invisible on the roentgenogram Rarely the cartilages become calcified and are demonstrable on the roentgenogram

Dislocation of the Ankle In the majority of fractures of the ankle, a dislocation of greater or lesser degree also occurs This is because the mortise of the joint is affected especially in Pott's fracture The foot is displaced laterally and reduction of the dislocation constitutes the first and most important maneuver in treatment With fractures of the posterior aspect of the lower end of the tibia the so called posterior malleolus, the astragalus is dislocated posteriorly In this instance also restoration of the proper weight bearing line of the lower extremity is essential Reduction of the dislocation is imperative and takes precedence over the correction of the fracture displacements Separation of the tibiofibular articulation may occur with or without associated fracture Re-

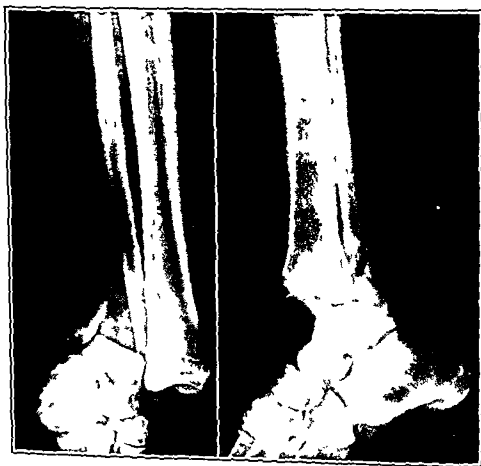


Fig 161 Fig 162

Fig 161 Dislocation of the Ankle and Tarsus There is dislocation of the ankle joint and the tarsal joints There is an oblique fracture of the lower end of the fibula with marked separation of the fragments

Fig 162 Dislocation of the Ankle and Tarsu Post reduction The same patient as in Figure 161 after reduction The reductions are not complete although the displacements are much less marked

coxa vara with decrease in the angle between the neck and the shaft The shaft of the femur undergoes atrophy The epiphyseal ossification center of the head of the femur on the side of the dislocation is delayed in appearing and is smaller than its fellow on the opposite side Shenton's line which normally curves smoothly along the superior margin of the obutrator foramen and the medial aspect of the neck of the femur, is altered and distorted The joint space is widened and the joint surfaces are irregular The ilium in the region above the acetabulum is flattened and decreased in size There is atrophy of the muscle tissues about the hip Untreated or unsuccessfully treated cases result in the formation of a false acetabulum above the true acetabulum Cases which are properly reduced early in life usually show increasing ossification of the head of the femur and eventual restoration to an essentially normal appearance While more commonly unilateral, bilateral dislocations occur in about 40 per cent of the cases The bilateral dislocations may be difficult of diagnosis particularly in early life Since early diagnosis and prompt therapy are of the utmost importance in order to prevent permanent deformity the possibility of the condition should be borne in mind and careful roentgen studies made in all cases

Dislocation of the Knee Dislocation of the knee is rare as very great force is required to disrupt the joint In complete dislocations

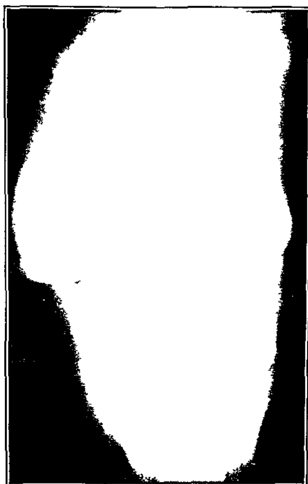


Fig 160 Dislocation of the Knee The patella is displaced laterally The soft tissues about the knee are markedly increased in density and thickness due to edema and hemorrhage The patient suffered a twisting injury of the knee

Dislocations of the Dorsolumbar Spine Dislocations of the dorsolumbar spine are rare and practically always are associated with fractures of the articular facets or pedicles Anterior displacement of the fifth lumbar vertebra with relation to the sacrum is termed spondylolisthesis and is discussed in detail separately

Dislocation of the Coccyx Anterior displacement of the coccyx or its segments occurs after a severe fall in a sitting position or a kick Hypermobility of the coccyx may be demonstrated by pressure on the coccyx during the making of the roentgen film The pressure may be applied externally or by a finger inserted into the rectum



Fig 163



Fig 164

Fig 163 Dislocation of the Cervical Spine There is posterior dislocation of the seventh cervical vertebra with relation to the sixth cervical vertebra The intervertebral space between the sixth and seventh cervical vertebræ is markedly narrowed There is a fracture of the articular processes of the sixth cervical vertebra

Fig 164 Dislocation of the Lumbar Spine There is marked lateral dislocation with displacement of the second lumbar vertebra to the right in relation to the first lumbar vertebra There is a fracture of the articular processes of the first lumbar vertebra

Dislocation of the Sacro-iliac Joint This requires very severe trauma and is most commonly seen in pedestrians struck by an automobile or train The entire side of the pelvis is displaced upward and outward There is simultaneous separation of the symphysis pubis or fracture of the pelvic bones

Separation of the Symphysis Pubis Separation of the symphysis pubis is common during or after pregnancy The conventional radiographs of the pelvis with the patient recumbent may not reveal the abnormality With the patient standing on one foot, the separation of the symphysis is manifested by the fact that one side of the pubis lies at a higher level than the other

duction is usually accomplished without difficulty True anteroposterior and lateral views are essential in the diagnosis of this lesion as oblique or semilateral views may mask the condition

Dislocation of the Tarsus In falls or severe twisting injuries, the tarsus may be dislocated The talo navicular joint may be involved or the cuboid be displaced Very marked swelling and deformity occur in tarsal dislocations Early diagnosis is of prime importance as operative measures become necessary unless reduction is effected promptly

Dislocation of the Toes While rare, this lesion is important as it causes interference with walking The anteroposterior and lateral view may not reveal the lesion because of overlapping, hence oblique views are necessary The phalanges are more commonly involved than the metatarsals

Dislocation of the Cervical Spine Dislocation of the first and second cervical vertebræ is very difficult of diagnosis The anteroposterior roentgenogram is made with the mouth open or the laminograph and reveals narrowing of the intervertebral spaces and overlapping of the bodies of the affected vertebræ In the lateral projection, rotary displacements are manifested by forward displacement of the posterior aspect of the first cervical vertebra with relation to the second or displacement of the odontoid process from its normal position in relation to the atlas Dislocations of the lower or middle cervical vertebræ are best demonstrated in the lateral projection There is anterior displacement of the affected vertebra in relation to the adjacent vertebral bodies The soft tissues of the retropharyngeal and retro tracheal spaces are increased in width and density and the tracheal and pharyngeal air spaces are usually narrowed Fracture of the pedicle or arch of the dislocated vertebra may be present also The roentgenologist has a definite responsibility in the demonstration of these lesions Accurate, early diagnosis may be a life saving measure Because of the patient's condition, proper positioning may be difficult or impossible unless both skill and care are used by the technician and roentgenologist In patients with short necks or broad shoulders the sixth and seventh cervical vertebræ may be obscured by overlying soft tissues However in practically every instance the entire cervical region may be visualized if the examination is performed carefully

Spontaneous Dislocation of the Atlas In children spontaneous dislocation of the atlas may occur after an inflammatory process in the nasopharyngeal region The patient complains of pain and stiffness of the neck usually of sudden onset in the absence of trauma The condition may occur in adults but is less common in the older age groups There is forward dislocation of the atlas with relation to the axis The dislocation is considered as being due to weakening of the transverse ligament of the atlas or distension of the bursa between the odontoid process and the anterior arch of the atlas There is painful torticollis and the patient finds it necessary to support his head with the hand It is essential that great care be used in the manipulation of the patient at the time of roentgen examination as sudden death may result from movement of the head with resultant pressure upon the medulla On the lateral roentgenogram the condition is characterized by diminution or obliteration of the space between the anterior arch of the atlas and the odontoid process The dens may be demineralized The distance between the ascending ramus of the jaw and the spine is increased and the posterior aspect of the arch of the atlas is anterior to the arch of the axis Fracture of the dens with forward displacement produces similar changes

fuse at seventeen to twenty years The styloid processes are developed from the epiphyseal centers In children, a trauma produces epiphyseal separations which correspond to the Colle's fracture of the adult A triangular fragment of bone is usually fractured at the lower end of the diaphysis of the radius and separates with the epiphysis Impaction is common Replacement is difficult as the epiphysis is relatively small and there is marked swelling of the soft tissues Proper reduction is important although growth is rarely interfered with if complete replacement is not achieved Fractures of the styloid process of the ulna may be overlooked unless it is borne in mind that the styloid process is an integral part of the epiphysis with no separate center of ossification

Elbow The epiphysis of the upper end of the radius is a disk-like structure The capsule of the elbow joint extends along the shaft of the radius below the epiphyseal line, hence separation of this epiphysis re-

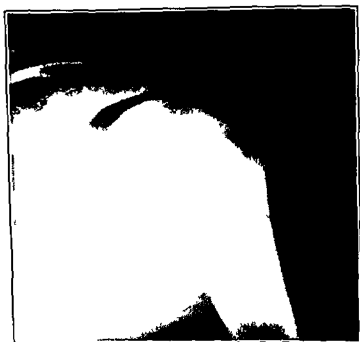


Fig 166 Separation of the Epiphysis of the Upper End of the Humerus

quires careful reduction The epiphysis of the upper end of the ulna lies at the extremity of the olecranon process and does not involve the articular surface Separation of this epiphysis is similar to a fracture of the olecranon

In the lower end of the humerus there are four ossification centers The capitulum appears at one to two years the medial epicondyle at five the trochlea at seven to eight, and the lateral epicondyle at ten to twelve years The trochlea and capitulum fuse with the lower end of the humerus while the internal epicondyle remains as a separate epiphysis until fusion takes place at about the age of eighteen years Supracondylar fracture usually occurs with epiphyseal separations in severe traumas to the elbow Dislocation of the elbow may also occur in children Interference with growth of the bones of the elbow is an important sequel of epiphyseal separation and must be guarded against by prompt and careful reduction Separation of the internal epicondyle epiphysis may result in wedging of the fragment within the joint with resultant dislocation during manipulation Removal of the fragment may be necessary

Dislocation of the Jaw (Temporo-mandibular Joint) This usually results from excessively wide opening of the mouth as in yawning or eating There is forward displacement of the condyle of the mandible with relation to the articular eminence Fracture of the condyle may result simultaneously or during reduction

EPIPHYSEAL SEPARATIONS

The epiphyses appear and fuse at relatively definite times in the case of each bone The development of the skeleton can be correlated with the calendar age and discrepancies in the relationships are important manifestations of physiologic disturbances In certain of the epiphyses double centers occur and are easily confused with fractures The epiphyseal line is weaker than the bone itself Before the time of union of the epiphysis a trauma applied to the region of the end of a long bone is



Fig 165 Separation of the Lower Epiphysis of the Radius The backward displacement of the lower epiphysis of the radius and the associated fracture of the lower end of the metaphysis of the radius are clearly demonstrated

most apt to cause an epiphyseal separation, a small portion of the adjacent diaphysis usually being fractured at the same time During adult life the bone is at its strongest and a similar trauma usually results in a dislocation In elderly people severe trauma as a rule results in fracture of the bone

Hand In the hand the epiphyseal lines are at the proximal ends of the phalanges and the first metacarpal in the remaining metacarpals, the epiphyses are at the distal ends The carpal bones are preformed in cartilage and ossify at the approximate rate of one each year, beginning with the capitate bone during the first year The pisiform is the last to ossify usually at about nine to ten years although it may not become calcified until the age of twelve years Injuries to the epiphyses of the hands are usually accompanied by fractures Wrestling fighting, and football are the common causes of these injuries

Wrist The lower end of the radius and ulna each have one epiphysis In the case of the radius the center appears at about two years of age The ulnar epiphysis becomes visible at about seven years They

a single center Epiphyseal separations are not uncommon and are often accompanied by a fracture of the adjacent portion of the metaphysis

Knee The upper end of the tibia has a single epiphysis which comprises not only the condyles of the tibia but also extends downward and anteriorly to comprise the tibial tubercle Separation of this portion of the epiphysis is commonly seen in football or jumping accidents in adolescents and is known as Osgood Schlatter s disease The epiphysis of the lower end of the femur develops from a single center and may be separated or involved in fractures of the lower end of the femur

Slipping of the Capital Femoral Epiphysis

Slipped capital femoral epiphysis is most frequently seen in males between the ages of twelve and fifteen years The involvement is usually unilateral although both sides may be involved The etiology is unknown A history of trauma can be elicited in many instances Interference with the blood supply of the head and neck of the femur are significant factors Many authorities are of the opinion that there is an endocrine basis for slipping of the upper femoral epiphysis, particularly in view of the fact that the lesion is in practically all instances accompanied by abnormalities of growth which are associated with an endocrine disorder As a rule, the disturbance of growth is the adiposo genital syndrome, a condition characterized by obesity and deficient gonadal development Less commonly there is a history of very rapid growth resulting in a tall, thin child In the obese type there appears to be a low level of sex hormone in the circulation, while in the tall, thin patients the reverse is true as regards the blood level of the growth hormone The growth hormone and the sex hormone influence the rate of proliferation of the cartilage cells in the epiphyseal plates with consequent changes in the thickness of the plates and the rate of growth of the skeleton The anterior pituitary hormone produces an increase in the thickness of the plates and accelerates skeletal growth The sex hormone retards the proliferation of cartilage cells with resultant decrease in the thickness of the plates and the rate of skeletal growth Estrogen acts through inhibition of the secretion of hormone of the anterior pituitary rather than by direct action on the epiphyseal plate and it is probable that testosterone acts in the same manner It appears that in the intervals between the activation of the gonads and the time that growth ceases the structure of the epiphyseal plate is dependent on the relative levels of the growth and sex hormones in the circulation Since slipping of the capital femoral epiphysis occurs in association with abnormalities of growth which are characterized by alterations in the level of these hormones it is believed that imbalance of the endocrines causes structural weakness in the epiphyseal plate Harris found that in epiphyseal separation the plane of cleavage passes through the third layer of the epiphyseal plate The growth hormone acts to decrease while the sex hormone increases the shearing strength of the epiphyseal plate, the changes being due to alterations in the thickness of the third layer of the epiphyseal plate These observations are of significance in affording an anatomical basis for slipping of the upper femoral epiphysis

Slipping of the capital femoral epiphysis is a serious lesion which may result in crippling and permanent deformity unless properly treated The condition is frequently overlooked or diagnosed erroneously Early

Shoulder Joint The superior humeral epiphysis ossifies from three centers, one for the head and one for each of the tuberosities These epiphyses begin to ossify from birth to two years They fuse into one and unite with the shaft at about eighteen years The epiphyseal line is angulated, the apex of the angle being directed upward Dislocation of this epiphysis is seen but very rarely and is usually associated with fracture of the diaphysis

The scapula ossifies from seven centers, two for the acromion, two for the coracoid, and one each for the body, the inferior angle and the medial border The clavicle has a single, linear epiphysis at its medial end This epiphysis appears at about the age of fifteen to seventeen years and unites at the age of twenty two



Fig 167 Separation of Tibial Epiphysis and Fracture of Lower End of the Diaphysis of the Tibia

Foot The epiphyses of the foot are similar to those of the hand and are at the proximal ends of all the phalanges and the first metatarsal, the epiphyses of the remaining metatarsals being at the distal ends There is usually an epiphysis at the proximal end of the fifth metatarsal If this epiphysis fails to unite as is not infrequently the case it remains as a small supernumerary bone and is called the os vesalius Comparison of the normal and injured sides is helpful as anomalies are frequently bilateral

Each of the tarsal bones ossifies from a single center They do not appear at yearly intervals as in the wrist The calcaneus is the only tarsal bone to present an epiphysis It is a curvilinear shadow at the postero inferior aspect of the bone and may rarely be separated by a sharp pull of the Achilles tendon

Ankle The lower end of the tibia has a single epiphysis which includes the internal malleolus The entire distal end of the fibula develops from

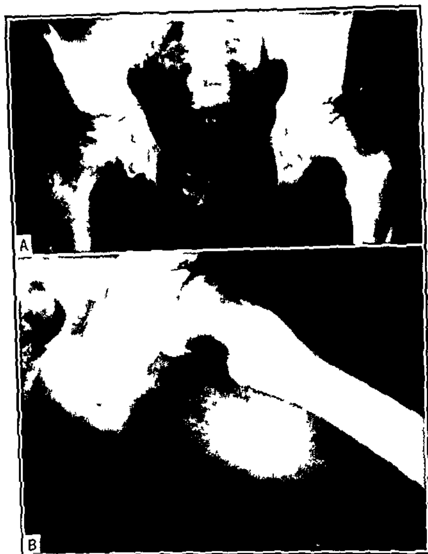


FIG 168 Slipped Femoral Epiphysis *A* Anteroposterior view There is slight downward displacement of the head of the left femur and the angle between the neck and the shaft is *abnormal* *B* Lateral view The displacement of the head of the femur is more clearly visualized than in the anteroposterior projection



FIG 169 Slipped Femoral Epiphysis *The epiphysis* at the upper end of the left femur is rotated and displaced the epiphyseal line is widened and the neck of the femur is short and broad There is osteoporosis of the bones of the left side of the pelvis and the left femur

diagnosis is imperative Otherwise operative procedures such as open reduction, arthrodesis, and the introduction of prostheses become necessary Clinically, the characteristic manifestations comprise limp and pain in the region of the hip The pain may radiate to the thigh, knee and leg The leg is rotated externally and there is limitation of flexion and internal rotation of the hip The diagnosis can be established with definiteness only by roentgen study The anteroposterior view is made with the patient supine, the lower extremities extended and the medial aspects of the feet in apposition The hips are maintained in a neutral position For the lateral view, the patient remains supine and the lower extremities are placed in the frog leg position with the hips flexed, abducted, and externally rotated The knees are flexed and the soles of the feet are placed in complete apposition The anteroposterior and lateral views are always taken with both hips on each film to permit of comparison of the two sides In the case of bilateral involvement the early phases of slipping are difficult to demonstrate and are frequently overlooked

The Normal Hip In infancy the neck of the femur forms an angle with the shaft which approaches 180 degrees The angle decreases with growth and weight bearing Ossification of the head begins between the third and seventh months, the ossification center being ovoid in shape After two years, it gradually assumes a hemispherical contour In the anteroposterior view after the age of three the head overhangs the neck so that prolongation of the superior neck line proximally transects a portion of the head The ossification center of the greater trochanter appears at about the age of three The juxtaepiphyseal portion of the neck after the first year appears slightly cupped in relation to the ovoid head This relationship is gradually reversed and by the age of twelve, the metaphyseal portion of the neck has become convex and the overlying head concave After the age of six years the medial contours of the head and neck describe a continuous arc in the anteroposterior view In the lateral view the anterior and posterior surfaces of the neck are concave and the head appears spherical The epiphyseal plate in infants is wide particularly in the lateral and medial aspects With increasing ossification the plate becomes thinner and the large peripheral gaps are gradually obliterated as the head envelops the neck The central portion of the capital epiphysis appears to fuse first Fusion of the head and neck occurs between the ages of seventeen and twenty in males and one to three years earlier in females The ossification center of the lesser trochanter appears between the ages of eight and thirteen The epiphyses of both trochanters usually fuse one to two years earlier than the epiphysis of the head and in later childhood and adolescence a capital epiphyseal plate wider than the epiphyseal plate of the greater trochanter suggests the possibility of a slipped capital epiphysis

Roentgen Manifestations In the typical advanced case the roentgen manifestations are characteristic and the diagnosis is made without difficulty As a rule, the head of the femur is displaced downward toward the lesser trochanter The femoral neck is short, broad, and flattened along its superior aspect The epiphyseal line is rough wide and irregular The femoral head is dense and may show slight mottling There is osteoporosis of the bones of the hip and the muscles of the hip and thigh show atrophy In long standing cases there is overgrowth of bone with spurring at the inferior aspect of the femur in the region of the junction of the head and neck Occasionally the displacement of the head is posteriorly

with increased growth of both the femur and the tibia This is particularly apt to occur in cases in which the fractured femur has been treated with metal plates and screws and has been considered due to the presence of the metallic foreign body Pain in the lower back and scoliosis may result from even mild degrees of asymmetry In the case of children with limbs of unequal length, the parents frequently object to operative procedures on the short limb, particularly since it is uncertain whether stimulation of growth can be attained Pease has attempted to obtain increased bone growth by increasing the blood supply to the cells of the epiphyseal cartilage by the insertion of metal or ivory screws in the metaphyseal region close to the cartilage plate and has succeeded in causing increase in the rate of growth after operation The amount of growth is variable and is dependent on many factors The acceleration in growth occurs for a period of two to three years after the operation, diminishing as the epiphyseal plate grows away from the region of the foreign material Secondary operations are necessary in some cases and appear to produce satisfactory results If the cartilage columns in the epiphyseal plates have undergone degeneration, the method is unsuccessful Ivory has been utilized in the past for stimulation of bone growth Metals must be used with caution as certain metallic substances, notably magnesium, are toxic

MEASUREMENT OF THE LENGTH OF THE BONES OF THE EXTREMITIES

It is frequently of great value to be able to measure the length of the extremities The measurements should be sufficiently accurate to record the comparative true lengths at any one examination and the precision of the measurements should remain constant for varying bone lengths in order to establish true increments of growth In the use of roentgen methods for the measurement of the length of the bones, the procedure must be simple, not too time consuming and of a sufficient degree of accuracy to permit of reliance on the determinations There should be sufficient detail on the roentgenograms to enable the observer to detect abnormalities of the bones and joints and study the epiphyses epiphyseal lines and soft tissues It is essential to be able to record any discrepancy in the lengths of the individual bones and to check this measurement as well as the total length of the limb by comparative measurements In most instances this data is required with reference to the bones of the lower extremities as the orthopedic surgeon wishes to know the exact length of the bones in cases in which epiphyseal arrest or other similar operation is indicated Surgical operation for arrest of the epiphysis is used in disturbances of epiphyseal growth which result in shortening of one extremity This occurs particularly in poliomyelitis although it may also develop after early fusion of an epiphysis due to local infection or trauma

The first satisfactory method for establishing exact bone measurements was introduced by Millwee in 1937 and was termed slit scanography The required apparatus consists of a roentgen table with a side rail and arm to hold a roentgen tube at a film target distance of 25 inches The tube is propelled along the length of the table by a motor driven gear In place of the usual circular cone, an adjustable slit is arranged between the tube and the patient As the tube is moved from one end of the table to the

and in this instance only the lateral view reveals the slipping With marked posterior slipping, the displacement is demonstrable in the anteroposterior roentgenogram, the head assuming a rounded form as it is projected through the overlying shadow of the neck of the femur Medial slipping is less common and when mild in degree is demonstrable only in the sagittal projection It is best demonstrated by the fact that prolongation of the line of the superior margin of the neck proximally does not transect the head

The early stages or slight degrees of slipping are extremely difficult to demonstrate and often present a serious problem in diagnosis In the anteroposterior views in the early stages of slipping there is a wide, irregularly-shaped epiphyseal plate with a sharply outlined metaphyseal border of the head There may be increased density and swelling of the soft tissues about the hip joint There is frequently generalized atrophy of the hip region Downward or medial slipping is determined by failure of the prolonged superior neck line to transect as much of the head as in the normal There may be periosteal proliferation at the inferior junction of the head and neck The lateral projections in the early stages show posterior or anterior displacement of the head, straightening of the anterior neck line and periosteal proliferation at the posterior junction of the head and neck At this stage, the diagnosis can be established only by very careful comparison with the normal

ARREST OF GROWTH OF THE EPIPHYSES

The arrest of epiphyseal growth to correct unequal length of the legs has become more satisfactory with increased experience Early operation is desirable Blount s operation, the use of removable staples is a definite technical improvement The determination of the optimum time for operation is difficult The discrepancy in length is best determined by use of teleroentgenography and slit scanography The ratio between the amount of shortening and the expected growth determines how much arrest is necessary The proper epiphysis to correct the discrepancy is chosen from standard tables giving the percentage of the limb growth contributed by each epiphysis In the case of the femur 80 per cent of the growth takes place at the lower epiphysis

LOCAL STIMULATION OF BONE GROWTH

Differences in the length of the limb in children have been treated by orthopedic surgeons with but indifferent results In the past, arrest of growth of the epiphysis has been accepted as the most feasible approach to the problem of correction of inequality in length It is known that increased vascularity in the region of or within the long bones produces acceleration of bone growth This occurs in congenital or acquired vascular anomalies local infectious processes cystic diseases tumors trauma and after surgical procedures on the bones or the soft tissues adjacent to the bones After osteomyelitis there may in the case of the femur be overgrowth of the corresponding tibia of two centimeters or more Stapling of the lower femoral epiphysis and the insertion of wires and metal pegs in the femur have resulted in stimulation and overgrowth of the subjacent tibia Fractures of the femur may cause a similar change

over the joints to prevent undue distortion The exposure factors which have proven most satisfactory are 42-inch distance, 100 m a , 48–55 kv For the ankles, the exposure time is one quarter second, knees one half second, and the hips, one second The films may be made also with the patient upright The position of the uppermost surfaces of the heads of the femurs with respect to the calibrated lines is noted and the relation of the internal malleolus of each ankle to the lines and lead markers is determined The difference between the two establishes the length of the limb If the part under measurement falls between the lines, a ruler may be used to determine the fractions The advantages of this method are the ease of making the roentgenograms, the accuracy of the measurements and the economy of films

A method used by Green and his co workers necessitates three separate exposures on a single roentgenogram The central ray is directed successively over each of the three joints and the three exposures are processed on a continuous film 14 inches in width and of any desired length up to a maximum of 42 inches The technical factors may be varied between exposures according to the thickness of the soft tissues of each part A target film distance of six feet is used constantly A tunnel for a long cassette measuring 14 × 44 inches has incorporated within it two sliding metal shields which permit an exposure to be made over any third of the film while the remaining portion is protected during the exposure The patient is placed on the cassette holder in the supine position with the hips level and the extremities parallel Straps are fastened over the lower thighs and ankles to maintain the limbs in proper position Three exposures are made in sequence with the tube centered over the hip the knee joint and the ankle the shileds being arranged to permit only exposure of the part desired The centering of the tube in each exposure is checked by a long metal marker placed horizontally at right angles to the side of the cassette with one end directed at the level of the joint to be exposed and extending over the cassette in such a manner as to cast a shadow on the film The other end indicates the position of the tube stand during the exposure In this manner the exact level of the focus is permanently recorded on the orthoroentgenogram The distance of 6 feet from the tube to the film eliminates inaccuracies which may result from imperfect centering The divergence of the rays from the tube produces considerable magnification even when a distance of 6 feet from tube to film is used However teleroentgenograms permit a fairly accurate estimate of the relative lengths of the two extremities When this method is used for serial measurements of growth, variable distortion ensues and becomes an important factor as the magnification increases with increase in growth The distances of the bones from the film cannot be definitely established in any given case However, the method gives a permanent record of the lengths of the bones of the lower extremities The fact that the entire lower extremities on both sides are visualized on a single film possesses certain important advantages which make the method of great practical value The disadvantages of the method are that most roentgen departments do not have the facilities for this type of examination

A modification termed spot orthoroentgenography has been suggested The technic is simple and the measurements are accurate The method can be utilized with any diagnostic roentgen equipment merely by the addition of a cassette which measures 14 inches by 36 inches and a brass

other, a beam of roentgen rays in the form of a narrow line traverses the patient The apparatus is so arranged that all of the film is protected by lead except the narrow strip at the time of exposure In this method the central beam of roentgen rays passes through the body and reaches the roentgen film from the same angle This results in a reduction of the effects of distortion

Mueller and Higgason modified this method by a system of spot scanography which has the advantages of being simpler, less time consuming and obviating the use of special apparatus The patient is placed on an ordinary roentgen table in the supine position with a 14 × 17 inch film under the thigh As it is advisable to include the hip and knee joint on the film, the roentgen film may be placed obliquely or lengthwise The roentgen tube is supplied with a small circular cone of sufficient size to include the hip or knee joint It is centered directly over the head of the femur at a 36 inch distance and the first exposure is made Without moving the patient or the film, the tube is then centered over the knee joint and a second exposure is made Another 14 × 17 inch film is placed under the leg to include the knee and ankle joints and in the same manner two small spot views are exposed one centering over the proximal end of the tibia and the other over the distal end of the tibia To measure the length of the femur a straight line is drawn from the tip of the femoral head to the medial lip of the internal condyle at the joint line In the case of the tibia a straight line is drawn from the medial tip of the internal condyle at the articular surface to the tip of the internal malleolus Comparison of the slit and spot methods of scanography shows that the two are of equal accuracy This method is simple accurate and requires no special apparatus It can be used for determining pelvic measurements and for measurement of the arm or other parts of the body

A common method has been to place a lead strip with calibrated perforations on the roentgenograms at the time of exposure The extremities are studied on large films which overlap each other in order to include the ankles knees and hips This has not proved satisfactory as it is not always possible to include the entire extremity on the films or improper exposure may fail to show a distinct outline of the individual joints A very large film may be used but this has the disadvantage that it is difficult to manipulate in the developing and drying processes A simplified method is suggested by Merrill A flat roentgen table and the Bucky diaphragm are used together with a specially constructed board The board is made up of two pieces of plywood three sixteenths of an inch thick 18 inches wide and 48 inches long One of the pieces of plywood is calibrated in inches or centimeters small holes being drilled in the board on each side at measured points and fine copper wire (26 gauge) drawn through the holes Lead numbers are placed on the wires, the numbers being arranged consecutively as on a ruler This surface of the board is given a coat of shellac and the top board is screwed over it The procedure for making the roentgenograms of the leg is simple The patient in a supine position is placed on the calibrated board which lies on the table top The tip of the internal malleolus should be placed near the number 4 lead marker Immobilization is essential and is obtained by sand bags at the feet and a strap across the thighs Three exposures are necessary The first film is an 8 × 10 inch centered over the ankles The second is a 10 × 12 inch centered over the knees and the third an 11 × 14 or 14 × 17 centered over the hips The centering must be accurately

Chapter

4

Infections

In Collaboration with IRVING A SHAUFFLR, M D

PYOGENIC OSTEOMYELITIS

THE terminology and classification of the various inflammatory diseases which occur in bones are still the subject of controversy The term osteo myelitis or osteitis indicates that there is involvement of the periosteum and the bone marrow as parts of the bone It is important to stress that the inflammation is confined to the interstitial connective tissue and the reactions of the bone tissue to the inflammatory process are secondary The disease may occur in an acute and a chronic form Both osteoclastic and osteoblastic activity develop as a reaction to the inflammatory process in bone The stage of the disease the virulence of the infection the resistance of the host, and other factors influence the extent and character of the changes

Acute hematogenous osteomyelitis is the most common form of the disease and occurs as a rule in childhood and adolescence The infection is metastatic in origin and usually begins in the marrow spaces of the metaphysis of the growing long bone, less often in the periosteum of the shaft or in the epiphysis During growth of the bones, the capillaries in the metaphysis form a hair pin curve at the point of junction of the connective tissue and the cartilage Retardation of the circulation in the capillaries favors the formation of bacterial emboli The process spreads to the diaphysis and the cortical compact bone to involve the inner layers of the periosteum The disease is characterized by necrosis and sequestration of large segments of the compact cortical layer, the formation of an involucrum, and extension of the process to the surface of the bone There may be perforation of the epiphyseal plate with involvement of the epiphysis the articular cartilage and the joint The lesion is usually caused by one of the staphylococci, usually staphylococcus aureus Localized acute osteomyelitis is the result of infection of a bone through the blood stream from the external surface as in a compound fracture, after tooth extraction or extension of an infectious process from the adjacent soft tissues or a pneumatic cavity of the skull such as the paranasal sinuses, mastoid or middle ear The process is usually rapidly progressive and is characterized by extensive osseous involvement There is resorption of bone periostitis formation of sequestra and reactive formation of new bone Chronic osteomyelitis is secondary to the acute variety in most instances It occurs particularly in cases in which the inflammatory

cylinder 26 inches long and $4\frac{1}{2}$ inches in diameter The cylinder is attached to a cone base with a bracket of a right angle type A vertical mark is painted on opposite sides of the cylinder to facilitate visual centering of the roentgenographic tube The patient lies supine on the cassette which is placed on top of the x ray table In order to determine the point at which the cylinder should be centered, each joint is flexed and a wax pencil mark is placed on the lateral aspect of the joint To prevent movement during the examination the patient is immobilized A pad is placed between the medial malleoli and the legs are tied closely together just above the malleoli with a length of 2 inch gauze bandage A cylinder of wood 1 inch in diameter is placed under the knee to provide firm support and maintain the knees at equidistance from the film A compression band is applied tightly over the thighs In the case of restless children, a second compression band may be utilized across the legs The brass cylinder is aligned with the mark over each joint and six exposures are made Roentgenograms of the left wrist and hand are taken to determine the bone age Due to the coning effect of the cylinder it is not necessary to block off the remaining portions of the film with lead The 40 inch target film distance simplifies the technic and the measurements are found to be as accurate as with 72 inch distance provided each joint is properly centered In order to avoid error, it is important that each joint be positioned in the center of the projected circle If the hip joint is shown at the extreme top of the upper circle and the ankle joint at the bottom of the lower circle an error of 1 3 per cent is possible The film itself may produce certain errors The average processing shrinkage of film 36 inches long is 0 25 cm Humidity expansion per 10 per cent of relative humidity is 0 11 per cent Thermal expansion per 10 degrees Fahrenheit is 0 06 per cent The possible errors due to the film characteristics are significant only when temperature and humidity are subject to extreme variation and can be eliminated by the use of a calibrated marker guide

fection is demonstrable after the seventh to tenth day Within the center of the reaction area one or more small areas of decalcification appear Bone destruction and loss of bone substance become manifest within about ten days and earlier in some instances Between the foci of de struction there are areas in which the trabeculæ are well preserved The margins of the destroyed areas are sharply spiculated, indicating that small segments of bone have temporarily resisted destruction Relatively early



Fig 170 Fig 171

Fig 170 Osteomyelitis of the Tibia There is extensive osteomyelitis involving practically the entire diaphysis of the tibia with both destructive and proliferative changes the former pre dominating There is periosteal elevation along the posterior aspect of the shaft of the tibia in the region of the junction of the upper and middle thirds The process does not involve the epiphyses

Fig 171 Osteomyelitis of the Humerus The diaphysis of the humerus shows muptiple areas of bone destruction advanced osteoporosis irregularity of outline thinning of the cortex and periosteal elevation The epiphysis of the upper end of the humerus and the shoulder joint space are not involved

in the course of the disease the periosteum is elevated This occurs in the region adjacent to the locus of the disease The elevation of the peri osteum is irregular in character Calcific deposition ensues in close rela tion to the cortex The calcific deposit varies in density but is less dense than the cortex and is irregular in contour A portion of the cortex which is undergoing sequestration presents no overlying calcification Sub periosteal calcification may be demonstrable at a considerable distance from the site of the infection and is of different character, being smooth in contour and of uniform density This first becomes evident in

process continues to be active due to the presence of a sequestrum which cannot be exfoliated spontaneously or involucrum formation During the chronic stage, the disease remains stationary or extends only very slowly and is characterized by the formation of large amounts of reactive bone

The primary chronic form of osteomyelitis is a chronic inflammation which has not been preceded by clinical manifestations of an acute onset The disease is divided into a simple and a granulomatous form The simple variety is caused by the pyogenic bacteria, most commonly the staphylococcus The granulomatous forms are caused by tuberculosis, typhoid, syphilis, the mycoses and occasionally leprosy

It must be stressed that in osteomyelitis the roentgen manifestations may be slow in developing and frequently lag far behind clinical evidences of the disease This delay may be as long as ten or more days, though usually it is about three to five days Therefore, in the presence of pain tenderness over the bone elevation of temperature, and a high white count, immediate surgical intervention may be indicated despite the presence of negative roentgen findings This is due to the fact that infection spreads along the medullary canal Prior to actual development of osseous destruction, the affected bone is of the same density as the normal bone and no change is distinguishable on the roentgenogram As the disease progresses, the cortex is penetrated at various points with islands of apparently uninvolved bone remaining between the areas of disease The process extends into the subperiosteal region with elevation of the periosteum The periosteum may be perforated and the pus escape to the surface Bone necrosis involves both cancellous and cortical bone and may be localized diffuse or scattered The bone destroyed by the infection is absorbed with resultant cavity formation Areas of necrosis in the cancellous bone are usually absorbed In the cortex areas of necrotic bone result in sequestration with the formation of bony fragments which are denser than normal The periosteum, endosteum and the cortex in the region of the infection are stimulated to new bone formation the so called involucrum The involucrum forms a new shaft continuous with the cortical portions of the bone in the uninvolved areas Superimposed layers of involucrum may cause marked increase in the width of the bone As sequestration occurs cavities result In children, these cavitations usually heal In adults, the cavities may persist for many years and result in discharging sinuses Persistent sinuses tend to develop epithelial linings and epidermoid carcinoma can originate in these areas The uninvolved bone in the affected region usually undergoes atrophy This usually disappears when use of the limb is re established Areas of low grade inflammation may persist for very long periods of time and often remain quiescent for months or years With lowering of the patient s resistance recurrence may result After complete healing, the bone structure may re establish itself so that no trace of the disease is demonstrable

Roentgen Manifestations Bone atrophy is usually the earliest roentgen finding and involves the cancellous portion and the cortex in the region of the epiphyseal line The periosteum is elevated in the case of the long bones Within a few days after the onset of an acute osteomyelitis a hazy non contrasty area of slightly increased density appears in the bone at the site of infection The bone trabeculations become indistinct Decalcification of bone trabeculæ at the site of in

Sequestra occur in practically all untreated or unsuccessfully treated cases of osteomyelitis A sequestrum may comprise a small irregular area, a linear fragment or a large segment of bone The sequestrum is manifested as an area of increased density In many instances there is a surrounding zone of diminished density The sequestrum may be resorbed or extruded In some instances, large sequestra serve as a framework for new bone formations Small sequestra lying in the soft tissues and in the process of being extruded may show decreased density Decalcification of a bone or all the bones of the affected extremity ensues after the disease has been present for a period of weeks or months Healing of osteomyelitis is manifested by disappearance of the swelling and reaction area The bone becomes clearly defined and sharp in detail

In chronic or long standing cases, the cortical bone shows marked increase in width and density with multiple irregularities of outline The medullary cavity is irregularly narrowed and in some areas may be completely obliterated Periosteal new bone formation is extensive The uninvolved bone shows markedly increased radiance due to atrophy, particularly in the segment distal to the site of involvement Sequestra become increasingly dense and the line of division between the involved and uninvolved portions becomes clearly demarcated Cavities and sinuses are visualized as areas of increased radiance and are usually irregularly ovoid or rounded in shape Pathologic fractures may occur Necrotic bone which has lost its connection with living reactive connective tissue as frequently occurs in suppurative inflammation cannot be resorbed In the early stages it is not possible to establish the diagnosis of sequestration either clinically or by roentgen methods After a short interval, the characteristic roentgen picture usually develops Sclerotic bone may form in sufficient amounts to encapsulate the sequestrated bone This is termed involucrum formation and is manifested roentgenographically by the presence of increase in the density of the affected segment of bone

Osteomyelitis of the Spine

Osteomyelitis of the spine is usually a fulminating infection and proves fatal in a high percentage of cases The disease is more common in males in the ratio of three to one The usual age of onset is in the third decade which is in contrast to the earlier occurrence of osteomyelitis in other parts of the body The most common infecting agent is the staphylococcus aureus the staphylococcus albus and streptococcus being less frequent The vertebræ are generally affected by metastases from a primary focus such as a boil, tooth infection, septic wound or other suppurative process The clinical manifestations are so protean and atypical that the condition may exist for a period of weeks months or longer before the diagnosis is established Because of the close anatomical relationship of the affected vertebræ to the segmental nerves the pleura, the peritoneum and other structures the clinical syndrome is extremely variable and the condition is confused with disease of the abdominal organs the thoracic viscera the central nervous system the hip the leg the knee and other portions of the body The lesion may affect any portion of the spine It occurs in all age periods from one year to seventy, the most common being between twenty and fifty years of age The disease may be acute subacute, or chronic However it frequently is not possible to determine the exact

the region directly adjacent to the elevated periosteum and is separated from the cortex by a narrow zone of radiance indicating that calcium is absent. The subperiosteal calcification may become confluent with the cortex in the later stages. These changes occur in any condition associated with elevation of the periosteum and do not signify necessarily that infection is present.

Proliferative changes manifested by new bone formation may develop relatively early, in some cases within ten to fourteen days after the onset. The involucrum may become so dense as to entirely replace the areas of



FIG 172 FIG 173

FIG 172 Osteomyelitis of the Femur. The shaft of the femur shows extensive scattered areas of destruction, irregularity of outline and periosteal thickening. The soft tissues of the thigh are increased in density.

FIG 173 Chronic Osteomyelitis. The patient had a fracture of the lower femur complicated by osteomyelitis. The increased density, irregularity of outline and new bone formation in the adjacent soft tissues are characteristic of chronic osteomyelitis.

destruction. Roentgen evidences of disease in the denser portions of the cortex may not be demonstrable until sequestrum has formed, which usually requires many days. The soft tissues show swelling, increased density and thickening. The process in the bone is usually confined to the diaphysis, being limited by the epiphyseal line. In the presence of a virulent infection, extension into the epiphysis and the joint is common. Rarely two or more bones may be involved. The areas of increased radiance in the bone are irregular, present poorly defined borders and may involve the entire shaft. Early roentgen studies indicate the site of origin and serial observations show the degree of virulence and progress of the disease. The process of healing can be estimated by the extent and sites of new bone formation and the reaction in the periosteum.

less marked than in tuberculosis In the lumbar region paravertebral abscesses usually do not develop as the pus descends along the psoas sheath on one side or bilaterally producing loss of definition or haziness of the psoas muscle shadows

Osteomyelitis of the thoracic spine may masquerade as primary pulmonary infection Localization of the lesion in the anterior portion of the pedicles and anterior surface of the lateral thoracic processes may be followed by purulent infection in the mediastinum and the paravertebral and extrapleural spaces with the formation of a mediastinal abscess The abscess may spread by direct contiguity or through the lymphatic channels and cause empyema, suppurative pericarditis, mediastinitis extrapleural abscess empyema, and pericarditis Superficial pointing of the abscess may occur very late or not at all The changes in the spine may be completely masked by the pulmonary manifestations, many of the cases being diagnosed as pneumonia, empyema, or carcinoma of the mediastinum

Osteomyelitis of the Skull

Osteomyelitis of the calvaria may occur in persons of any age but is most common in those between twenty and thirty years It appears to be less common in older individuals The greatest number of cases follow disease of the frontal sinus A few are subsequent to infection of the ethmoid cells the antrum the sphenoidal sinus, and the mastoid Skull trauma with extension of infection from the scalp and metastatic or hematogenous infection are also causes of osteomyelitis of the bones of the skull Any infection such as the common cold scarlet fever, and influenza may be complicated by osteomyelitis Swimming, operations on the sinuses, and skull operations have been responsible in some cases The process spreads via the diploe Early in the disease there is engorgement of the vessels thrombophlebitis, and pus in the interspaces of the diploe Both tables are affected, as a rule the inner to a greater extent than the outer If the lesion breaks through the inner table an extradural or brain abscess results Spread through the outer table produces subperiosteal abscess and swelling of the tissues of the scalp and forehead In some instances the disease begins without any antecedent history of sinusitis Systemic symptoms and fever may be absent, mild or very severe

Roentgen Findings In the early, acute stages osteomyelitis of the bones of the skull may produce no roentgen signs for periods as long as two to three weeks or more This cannot be emphasized too strongly as delay in the institution of therapeutic measures until the definite demonstration of x ray changes may result in the development of serious complications Areas of increased radiance and rarefaction of the bone are the first findings on the roentgenogram The rarefied areas stud the involved bone with segments of normal bone interspersed Coalescent spread results in the formation of large segments of bone destruction Slight rarefaction of the bone precedes the areas of definite involvement indicating advancement of the infection If the process is disseminated via the diploic vessels areas of rarefaction which have the appearance of small venous lakes appear and new foci of bone destruction form at a considerable distance from the original site with normal bone intervening In children the condition is apt to be limited in its advance by the sutures In adults there are no natural barriers and the process may produce very

stage of the disease or its duration on the basis of either the clinical manifestations or the roentgen findings There is moderate elevation of the white cell count It some instances the causative organism can be cultured from the blood stream The vertebræ are particularly susceptible to infection because of the anatomy and structure of the bone The cancellous portion of the bone contains an extensive sinusoidal venous system which favors stasis and bacterial proliferation Organisms which have gained entrance to the vertebra multiply rapidly and cause destruction of the bone trabeculæ The vertebral and intervertebral structures weaken and collapse Herniation of the nucleus pulposus may ensue The neural arch and the spinous and transverse processes are highly susceptible and frequently show extensive involvement

ROENTGEN MANIFESTATIONS The early manifestations of the disease comprise a typical osteoclastic reaction In most cases there occurs simultaneously with the necrosis and dissolution of bone a variable degree of production of new bone This is very important in differentiating the lesion from tuberculosis The disease is characterized also by extensive destruction of the intervertebral disc, apparently due to direct extension of the infection The disc may herniate into the diseased vertebra The infection spreads from the vertebra to the disc Involvement of the adjacent soft tissues is less frequent than in tuberculosis In tuberculosis the lesion extends along the ligaments producing large paravertebral abscesses While paravertebral abscess occurs occasionally in pyogenic osteomyelitis extension of the disease to adjacent segments of the spine is by direct extension across the disc spaces The pus does not collect under the anterior ligaments to produce erosion as in tuberculosis, actinomycosis and other granulomatous infections Extension into the psoas sheath is frequent because of the anatomical attachment of the psoas structures In the early stages of the disease there may be narrowing of the disc space This is similar to the change which occurs in tuberculosis and frequently results in confusion in diagnosis However the bone change in early pyogenic osteomyelitis comprises rarefaction with involvement of the inferior or superior lips of the body anteriorly As the process advances the areas of rarefaction increase in size The degree of rarefaction and demineralization is more marked and the extent and rapidity of the bone destruction are much greater than in tuberculosis The important diagnostic feature in osteomyelitis is the early appearance of bone reaction and proliferation in some instances definite changes being present within four to six weeks after the onset of the disease In tuberculosis of the spine this manifestation is extremely rare Involvement of the spinous process the transverse process or a portion of the neural arch without evidence of disease in the body of the vertebra strongly favors the presence of osteomyelitis There is usually a lesser degree of deformity and collapse of the disc in osteomyelitis than in tuberculosis most probably due to the fact that the latter disease is more chronic and is usually associated with more extensive disruption of the disc and vertebra Modern drug therapy with the sulfonamides and antibiotics has resulted in marked alteration of the picture in both osteomyelitis and tuberculosis

In osteomyelitis the soft tissue changes are minimal or entirely absent and are dependent upon which segment of the spine is involved In the cervical region there is loss of definition of the intermuscular lines and anterior displacement of the pharyngeal and tracheal air shadows The thoracic region is apt to show a globular paravertebral swelling This is

The margins of the lesion are smooth and sharply defined There is no periosteal reaction Small central sequestra may occur Swelling is absent The course is benign These features differentiate it from eosinophilic granuloma, metastatic carcinoma, and other similar osteolytic lesions which may produce somewhat similar roentgenographic manifestations

Chronic Osteomyelitic Changes in Association with Neurotrophic Disturbances Neurotrophic Changes in Bones

The bones not infrequently show changes which are considered as being neurotrophic in origin The manifestations comprise atrophy and areas of rarefaction with extensive absorption of the bone in the affected areas In



FIG 175 Erosive Defects in Paraplegia There is advanced destruction of the lateral margins of the iliums and the right ischium The patient was a paraplegic

many instances, the bone lesions are accompanied by trophic ulcers of the adjacent soft parts The changes may involve both the tubular and flat bones and have been described in tabes syringomyelia leprosy post-traumatic paraplegia and similar conditions The earliest roentgen change is osteoporosis Subsequently there is localized or widespread destruction of bone The areas of destruction present irregular poorly defined borders with no evidences of sclerosis A characteristic alteration associated with involvement of the heads of the metatarsals and the phalanges is a decrease in the calibre of these bones with the production of a pencil point type of deformity The periosteum may show thickening and proliferative activity along the shaft of the involved segment The distal segment of the bone gradually undergoes absorption and the cortex becomes thin or disappears The medullary portion is absorbed

extensive involvements As healing takes place the rarefaction of the bone disappears gradually Sequestra may form and are at times very difficult to identify as they closely resemble areas of normal bone Bone degeneration begins at the periphery or within the areas of destruction and healing in irregular patches may take place Irregularities of outline of the bone and mottled areas of density often persist for long periods after the lesion has healed clinically

DIFFERENTIAL DIAGNOSIS The differential diagnosis of osteomyelitis is concerned particularly with syphilis, tuberculosis and sarcoma In syphilis there is usually marked bone proliferation with little or no *destruction The changes involve both the cortex and periosteum* The process is multiple and affects several bones simultaneously Sequestra are not produced except in the calvaria The adjacent soft tissues are



FIG 174 Osteomyelitis of the Skull

unaffected Bone tuberculosis usually involves the bones of the hands and feet although in very young children the long bones may be involved The process is primarily destructive with little or no proliferation Sequestra may occur about the joints rarely in the diaphysis The periosteal new bone formation is usually laminated The process invades the epiphyses joints and adjacent bones Negro and Chinese children are among those most frequently affected Ewing s sarcoma may in certain instances be extremely difficult to differentiate from early osteomyelitis Ewing s tumor is usually single does not form sequestra and the new bone is apt to be formed in successive layers termed onion skin layers or lamellations In osteogenic sarcoma the new bone formation is formed in irregularly radiating spicules and no cavities or sequestra are present The process is limited to one bone and there is no linear cortical thickening Brodie s chronic abscess of bone is a localized osteomyelitis characterized by a solitary area of bone destruction in the cancellous portion of the bone

and poorly defined There is complete absence of bone within the affected area Surrounding the area of increased radiance, there is a zone of increased density which has indefinite margins and gradually merges with the adjacent normal bone The overlying soft tissues present no evidences of swelling, distortion or alterations in density The sclerosis surrounding the focus of infection apparently does not wall off the area of infection Rather it appears that early and extensive bone production occurs as a consequence of the low grade chronic infection and is a manifestation of the unsuccessful attempt of the bone to heal the process

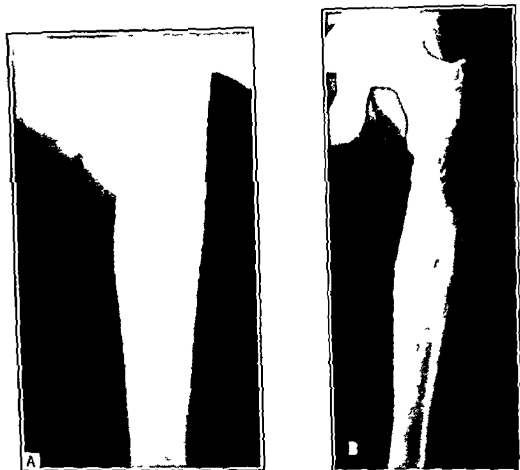


FIG 1/7 Sclerosing Osteomyelitis *A* Prior to operation The upper third of the shaft of the femur reveals increased density and marked cortical *thickening the* characteristic manifestations of sclerosing osteomyelitis of the Garre type *B* After operation The rounded area of radiance and the small shadow of metallic density *in the medullary* aspect of the femur represent the operative defect and a small fragment of a drill broken by the extremely hard bone during the operation

The Early Diagnosis of Osteomyelitis

Roentgen study is of the utmost importance in establishing the diagnosis of osteomyelitis in the early stages The value of this cannot be overemphasized as it permits of the *institution* of antibiotic therapy and chemotherapy prior to the onset of bone *destruction* In many instances the diagnosis can be made on the second or third day after the onset of symptoms It is helpful in making the roentgenogram to use a special technic with low kilovoltage increased *focus* skin distance, and short object film distance Immobilization of the limb during the time of the x ray exposure is absolutely *necessary as* the least motion causes blurring

and the cortical walls are approximated The osteoporosis and the ab sorption may progress until the affected bone disappears practically completely The process tends to undergo spontaneous arrest It is the concensus that the bone changes termed neurotrophic in origin are most probably due to infection of the contiguous tissues with a low grade osteomyelitis secondary to the chronic infectious process in the soft tissues

Sclerosing Osteomyelitis of Garre

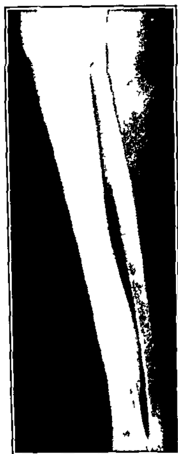


FIG 176 Sclerosing Osteo myelitis of the Tibia There is an area of increased density in volving the tibia in the region of the junction of the lower and middle thirds of the shaft There is new bone formation along the cortex the new bone being in layers parallel to the shaft The medullary cavity is partially obliterated in this re gion The roentgen diagnosis of sclerosing osteomyelitis was confirmed at operation

A rare form of osteomyelitis known as scle rosing osteomyelitis of Garre often is not diag nosed until persistence of symptoms leads to exploratory operation Localized pain, tender ness, and the absence of constitutional symp toms and significant laboratory findings should suggest the true nature of the condition Histo pathological examination discloses sclerotic bone with mild inflammatory reaction The roentgen appearance of chronic sclerosing osteomyelitis is usually typical The affected portion of the bone shows an area of increased density with cortical thickening and oblitera tion or narrowing of the marrow canal The affected area may be smooth or irregular in outline The periosteum is not elevated Rarefaction usually does not occur, although in long standing cases small, irregular areas of increased radiance may be present within the dense zones There are no changes in the over-lying soft tissues Ewing s endothelioma and osteogenic sarcoma may produce closely similar changes Paget s disease may cause confusion in diagnosis Paget's disease usually involves many bones and is characterized by greater central density and an increase in serum phosphatase Syphilis is excluded by the absence of a history of luetic infection and a negative serologic test Syphilitic periostitis usually involves a larger area than in scle rosing osteomyelitis Osteoid osteoma may produce changes very similar to those in sclerosing osteomyelitis of Garre and presents a difficult problem in differential diagnosis

Brodie's Abscess

Brodie s abscess is a form of chronic osteomyelitis The disease commonly occurs in the shaft of the long bones The inflammatory process is self limited It involves the marrow the compact corti cal bone and the periosteum Roentgenographically the lesion is manifested by an area of destruction with extensive formation of new bone The defect in the bone is localized and its margins are irregular

from that which occurs in acute osteomyelitis However there is a different distribution as the changes cross the joints and often affect an entire extremity rather than being confined to a single bone

Osteomyelitis in Infants

Osteomyelitis in infants may present important anatomical and clinical differences from the disease in older children and adolescents The prognosis and therapy may be affected by these variations The lesion is apt to occur in infancy subsequent to the common respiratory diseases and in umbilical and skin infections In many instances no causative primary infection can be determined The streptococcus is the most common pathogenic agent although staphylococcus aureus, gonococcus, penumococcus, B coli and pyocyaneus and other bacteria may cause the disease There are several anatomic factors which contribute to produce variations in the nature of the disease in infancy The bone presents a more spongy texture, the vascular spaces are larger, and the cortex is thinner than in older individuals permitting a free communication between the marrow and the subperiosteal spaces The most common mode of infection is through the capillaries in the juxta epiphyseal region of the metaphysis The cortex is very thin and the process is manifested by predominantly periosteal changes the medullary cavity of the diaphysis being involved only rarely The periosteum may rupture with the formation of soft tissue abscesses There is little if any bone destruction and sequestration rarely occurs Osteomyelitis in infants may occasionally develop primarily in the epiphysis Pyarthrosis is a common complication The infection may reach the joint by direct spread from the metaphysis or through the adjoining epiphyseal cartilage As the cartilage is resistant to infection epiphysiolysis between the metaphysis and the epiphyseal cartilage may occur In many instances the infection is sufficiently virulent to break through the cartilage into the epiphysis and extend into the joint Joints in which the epiphyseal cartilage and a portion of the metaphysis lie within the joint capsule, as for example the proximal and distal ends of the humerus and the femur become involved by direct spread of the process through the periosteum of the metaphysis The cartilage of the joint is relatively resistant to low grade infection In virulent infections the cartilage and epiphysis are destroyed with resultant deformities and arrests of growth Metastatic involvement of other bones is common Metastatic soft tissue abscesses may occur particularly during the stage of septicemia In infants healing occurs rapidly after surgical therapy or spontaneous regression The necrotic bone is small in amount and is absorbed promptly Persistent sequestra are rare There is rapid formation of new bone manifested by the development of subperiosteal new bone within a week after onset of the infection

Clinical Manifestations The clinical course in the *infant* is different than in other age groups The onset is gradual The first complaint usually comprises swelling and loss of function of the involved extremity Systemic manifestations are slight and may consist only of a low grade fever and slight elevation of the white count The condition is frequently confused with suppurative arthritis cellulitis soft tissue abscess scurvy and congenital syphilis In some cases there *may* be a more acute onset with severe generalized manifestations Rapid healing and regression are usual in infants and the disease may run its course in three to eight weeks

and may obliterate the slight changes in the roentgenogram on the basis of which the diagnosis may rest Osseous changes may not be demonstrable until the fifth to tenth day of the disease However, changes in the soft tissues adjacent to the affected bones become visible within a matter of hours after the onset of bone involvement Routine roentgenograms are often of little value in determining the presence of soft tissue changes because the detail of the soft parts is poorly visualized However, roentgenograms made for bone detail afford very important data when studied under a spotlight or similar strong source of illumination

The most significant early change is roughening of the sharp line of demarcation which is normally present between the subcutaneous shadow and the muscle bundles A second important early abnormality is blurring or obliteration of the intermuscular cleavage planes In the muscular and subcutaneous regions, the normal appearance becomes altered by the presence of multiple transverse, irregular lines of increased density extending from the muscle borders into the subcutaneous shadows The lines are not straight or regular in length and width rather produce a criss cross pattern which suggests that they may be vascular and lymphatic channels which have become widened and tortuous The intermuscular planes become obliterated and increased in density rather than forming sharp, clear cut lines The manifestations vary in extent and character in accordance with the severity and duration of the disease The abnormalities involve the soft tissues about the affected bone and do not extend beyond the adjacent articulations unless there is involvement of the joints With the development of periosteal reaction and a break in the cortex the abnormalities in the soft parts tend to regress, but may persist in modified form throughout the chronic stage of the disease

The principal factors in the production of the soft tissue changes are increase of the pressure and vascularity within the marrow cavity in consequence of the infection with transmission of these manifestations to the Haversian systems and the surrounding soft tissues There is not only dilatation of the vessels but also the formation of many others which previously comprised merely potential channels with edema of the muscles and surrounding stroma The degree of the changes is proportional to the extent and severity of the disease process The toxic products of the infection may play a part in the production of the soft tissue abnormalities There is local obliteration of the trabeculæ manifested by slight haziness at the site of the lesion The soft tissue changes are not uniformly present They are not entirely characteristic and in themselves do not permit of the establishment of a definite diagnosis However they are present for a considerable period before the bone manifestations develop and afford an important clue to the diagnosis

In differential diagnosis it is necessary to consider acute sprains abscesses of the soft tissues tumors of bone and soft tissue blood dyscrasias with extravasation and edema The principal aid in differential diagnosis is the distribution and extent of the changes In a sprain, there is localized swelling of the surrounding tissues but rarely extension along the entire shaft of the bone Soft tissue abscesses similarly are localized in extent Tumors present a soft tissue density which varies widely in size There is minimal extension of the changes in the stroma with no obliteration of the normal lines of demarcation except in the involved area Extravasation of blood or edema produces a picture which cannot be distinguished

lished prior to the administration of penicillin it often becomes impossible to make the diagnosis by roentgen study alone Early and accurate roentgen diagnosis is of the utmost importance If therapy is not instituted prior to the stage of bone destruction, its effectiveness is reduced This is particularly true in the presence of sequestra

OSTEITIS PUBIS

Osteitis pubis is a self limiting, non suppurative infection of bone which usually begins at the region of the symphysis The process extends to the pubic bones and in some cases also involves the ischial tuberosity



FIG 178 Osteitis Pubis There is advanced osteitis involving the pubic bones with rarefaction and irregularity of outline of the body of the pubis bilaterally and marked widening of the symphysis pubis The condition developed subsequent to prostatectomy The bladder contains sodium iodide solution

It has been termed periostitis pubis and osteochondritis pubis The lesion usually develops as a post-operative complication in urologic surgery It is essential that the condition be differentiated from osteomyelitis of the pubic bone because of the marked differences in therapy and prognosis It has frequently been confused with metastatic carcinoma and other destructive lesions Because of errors in diagnosis patients have been subjected unnecessarily to resection of the involved area wiring of the symphysis chordotomy and other serious and mutilating operations Roentgen therapy has proven to be the ideal mode of treatment which makes the condition of unusual interest to the radiologist The disease was first described by Beer in 1924 and since that time increasing numbers of cases have been reported It has been known to develop after supra

Roentgen Manifestations The initial roentgen manifestation comprises soft tissue swelling This is an indication that the process has involved the adjacent joint or soft tissues by extension through the periosteum The earliest bone change is an area of destruction in the metaphysis adjacent to the epiphyseal line The alterations are similar to those which occur in epiphysitis The periosteum of the diaphysis is elevated The joint space may be widened Before the epiphysis becomes ossified, it is not possible to determine whether or not it is affected If the epiphysis has been decalcified and destroyed it is necessary to determine by serial roentgenograms and comparative studies of the contralateral side whether the epiphysis is viable Partial or complete separation of the epiphysis frequently occurs and is an indication of infection involving the epiphyseal cartilage Healing is manifested by rapid subperiosteal formation of new bone and decrease or disappearance of the bony defects Subsequently there is restoration of the bone trabeculae Sequestrum formations are rare and frequently are absorbed spontaneously Restoration of normal bone growth is dependent on the extent of the epiphyseal destruction

The course of the disease is best observed by serial roentgen studies In many instances there is complete healing within two months, although minimal deformities may persist for many weeks or months In some cases there is delay of epiphyseal growth or failure of development of the epiphysis with resultant arrest of development Sclerosis and other signs of chronic osteomyelitis are rare The prognosis is good in infants who survive the original septicemia The progress of the local lesion is usually excellent The anatomical features in the infant permit of early natural decompression of the lesion The metaphyseal abscess drains into the subperiosteal space or the neighboring joint In infants with primary suppurative arthritis the possibility that the disease originated as osteomyelitis must be considered The roentgen examination is invaluable in the detection of the disease to follow the progress and in the evaluation of the end results

The Effects of Antibiotic Therapy in Osteomyelitis

The roentgen manifestations of osteomyelitis may be markedly altered by modern methods of therapy particularly the administration of the antibiotics In the patient with well developed osteomyelitis the drugs may mask the symptoms although bone destruction and sequestration continue unchecked Frequent roentgen examinations are essential to observe the progress of the disease In some instances, osteomyelitis develops during the administration of the sulfonamides or antibiotics Clinical symptoms are usually minimal or entirely absent and soft tissue swelling and reaction areas may not be present Local decalcification and bone destruction are the prominent manifestations The lesion lacks the important diagnostic features of osteomyelitis and may be diagnosed erroneously as neoplasm or leukemic infiltration The administration of penicillin to a patient with early osteomyelitis may abort the disease following the stage of soft tissue swelling and bone reaction and prevent progression to the stage of decalcification and bone destruction After the administration of the antibiotic drugs the reaction area disappears rapidly, the subperiosteal calcification may not develop, and sequestration may not ensue If the roentgen diagnosis of osteomyelitis is not estab

separation of the symphysis pubis Extensive decalcification occurs There is no sequestrum formation In rare instances the periosteum and segments of the underlying bone become separated by the pull of the muscles producing an appearance of false sequestration (3) Regenerative stage After a period of two to five months recalcification ensues and progresses slowly The affected bone is restored to normal after six to twelve months In the early periods of this stage of the disease, areas of rarefaction are intermingled with zones of beginning reossification The osteoporosis gradually disappears and solid bone with normal trabeculæ supervenes The symphysis may remain widened More frequently it becomes narrowed and in many instances it is completely obliterated In the stage of complete healing there is osteosclerosis of the involved bone with increased density in the region of the symphysis and multiple spur formations at the sites of the muscle attachments due to periosteal new bone formation The spurs are particularly prominent along the inferior rami of the pubis and the tuberosities of the ischium

Differential Diagnosis Osteomyelitis is the most difficult problem in differential diagnosis There is no fever or sequestrum formation in osteitis pubis In osteomyelitis there is a history of direct trauma to the pubis such as a fall or being thrown against a saddle while riding horseback or an indirect trauma to the knee leg or foot Mild forms of osteomyelitis are easily confused with osteitis pubis The differentiation is made by the presence of suppuration which develops in osteomyelitis and is absent in osteitis pubis Metastatic carcinoma must also be considered in differential diagnosis, particularly since the operation may have been a prostatectomy for primary carcinoma of the prostate Metastases from carcinoma of the prostate usually produce osteoblastic rather than osteolytic changes It is also necessary to consider tuberculosis and syphilis of the pubic bones If a sinus is present, roentgen studies after the injection of an opaque medium may be helpful in establishing the diagnosis

Prognosis The prognosis is good Deaths have been reported These occurred in patients with severe purulent infection of the pelvic structures and terminal septicemia The period of disability varies from three to six months and is followed by a stage of limited capacity which may last six to twelve months Practically every case has shown complete recovery after two years

Etiology and Pathogenesis There have been many theories advanced to explain the genesis of osteitis pubis (1) The theory of infection of the traumatized os pubis Many observers consider it necessary that there be injury to the periosteum Others feel that the important etiologic factor is injury to the periosteum of the os pubis or its nutrient vessels with spillage of the urine or septic material on the bone The injury to the periosteum may result from avulsion of the fibers of the rectus muscles at the point of insertion with resultant tearing of the pubic attachment of the transversalis fascia or other puboprostatic ligaments Pressure of retractors or drainage tubes puncture of the needle used for local anesthesia and interference with the blood supply are considered to be contributory factors Direct extension of a septic process through denuded areas of periosteum does not appear to occur An important factor is injury to the capsule of the prostate with leakage of septic material The condition is rare in females as the fascial planes are modified and the lymphatic drainage is chiefly by way of the broad ligaments The infection causes a low grade inflammation and sequestration is absent It

pubic prostatectomy, retropubic prostatectomy transurethral resection perineal prostatectomy, cystectomy pelvic ureterolithotomy, abdomino perineal resection of the sigmoid, pyelonephritis, a single external trauma to the symphysis and other conditions

Symptoms In some instances, the disease is mild and the condition is not detected until roentgen study of the pelvis is made As a rule pain is the predominant symptom The pain is in the region of the symphysis pubis and in some cases the onset is sudden and of great severity The pain radiates to the perineum the thighs, and Scarpa s triangle It is aggravated by movement of the trunk and thighs The pain is due to the pull of the muscles at the sites of attachment to the diseased bones and in many instances becomes excruciating in character As the disease extends from the symphysis pubis to involve the pubic rami and the tuberosities of the ischium the attachments of additional groups of muscles become affected and the symptoms and disability increase There may be marked spasm of the bladder and rectum The duration of the pain is variable In mild cases it may last for only three to four weeks In more severe and chronic cases it persists as long as four to twelve months Even after the subsidence of the acute manifestations there may be recurrent attacks during many months The patient may have a gait which resembles that in congenital dislocation of the hip There may be great difficulty in changing from the standing to the sitting or recumbent positions In severe cases the patient must use a crutch or a cane due to the severe atrophy of the muscles

Physical Signs There are few physical signs At the onset there is slight or moderate swelling over the symphysis with localized tenderness There are no manifestations of local inflammation or accumulation of pus The temperature and pulse usually remain normal or are elevated only slightly Extension to the pubic rami and the ischium is associated with tenderness on palpation or pressure in these regions There is no swelling of the lymph nodes as in osteomyelitis of the pubic bones As a rule the operative wound heals well, although in some cases a sinus persists If a sinus is present probing results in exposure of bare bone There may be thickening and irregularity of the bones due to hypertrophy in the late stages of the disease There is atrophy of the muscles of the thigh particularly the adductor muscles

Roentgen Manifestations In most cases there is a characteristic roentgen picture It must be stressed that an interval of days or weeks must elapse before the manifestations develop Once the disease begins there is progressive involvement of the bones There is a self limiting peak and after this has been attained the process undergoes reversal with gradual regeneration of the bone The disease is divided into three stages (1) Initial stage This period may extend for one to seven weeks after the onset of clinical symptoms and four to twelve weeks after the operation The roentgen examination is usually negative or there may be evidence of slight fraying of the margins of the pubic bones (2) Destructive stage The first signs of destruction usually appear in the medial aspect of the bodies of the pubic bones The involvement extends gradually to the rami and tuberosities of the ischium Roentgen study at this time shows roughening and fraying of the periosteum with a wooly motheaten rarefaction of the affected segments of bone The changes are most marked in the region of the symphysis pubis In the presence of marked involvement of the cartilage there is apparent widening and

a few weeks or months and the period of disability is reduced Complete re ossification of the bone develops within two years

ADDITIONAL READING

ABRAMS M SEDLETZKY I and STEARNS D B Osteitis Pubis N E J Med *240* 637 1949
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OSTEITIS OF THE SPINE

Osteitis of the spine is a rare complication of prostatic surgery and occurs much less frequently than osteitis of the pubis Metastatic cells from carcinoma of the prostate may be spread through the prostatic venous plexus to the vertebral column and infected material from the prostate may be spread in the same manner A small number of cases of osteitis of the spine subsequent to prostatic resection have been recorded in the literature The changes are similar to those which occur in osteitis pubis two or more vertebrae usually being involved The lesion undergoes gradual healing spontaneously and the progress of the repair is demonstrable by successive roentgen examinations Complete healing is manifested by fixation of the involved segments of the spine with calcification and ossification The development of these changes differentiates the lesion from metastatic carcinoma of the prostate

TUBERCULOUS OSTEITIS TUBERCULOUS OSTEOMYELITIS

Tuberculous infection of the bones is rare in adults, usually being a disease of the young The infection is hematogenous in origin and most commonly involves the long bones, particularly in the region of the hip, knee and ankle and the vertebral bodies The process may be proliferative or consist of destruction of bone or caries The lesion has a tendency to spread to the adjacent soft tissues and joint Reparative formation of new bone occurs characteristically in tuberculous osteitis of the long bones, particularly the phalanges the clavicle and the mandible. In some instances the new bone formation is very extensive The affected bones present a spindle shaped or fusiform swelling This type of change appears to be limited to bones which are under bending or shearing stresses and indicates that the formation of new bone is due to mechanical forces as a compensation for the loss of bony substance, stimulation by toxins playing only a minor role Since the manifestations of tuberculous osteomyelitis vary widely in the different forms of the disease it is necessary to discuss each variety separately

Disseminated Cystic Tuberculosis

Disseminated tuberculosis of bone is characterized by several significant features The lesions are frequent in the small bones of the hands and feet and are associated with a tendency to the formation of cyst-like defects with little or no proliferative reaction The patient usually presents a negative tuberculin test and tubercle bacilli cannot be demonstrated Other important aspects comprise absence of caseation, a

is not definitely known which bacteria cause the disease Practically every organism found in the urine has been suggested as the pathogenic agent An important factor is that urine acts as an irritant and may lower the resistance of the bones to infection

(2) The theory of acute atrophy or aseptic necrosis of the pubic bone Experimental attempts to reproduce the condition in rabbits by a combination of trauma and infection have not proven successful There is evidence that extensive decalcification of a large area of bone in the pubic and ischial rami takes place *It is not usual for an infection to cross the* cartilaginous symphysis and extend an equal distance to each side of a joint Healing of osteitis pubis results in normal trabecular restoration of the bone while periostitis is characterized by deposition of new bone with destruction of the trabeculæ Osteomyelitis is associated with sequestrum formation and destruction of bone trabeculæ According to some observers, the condition constitutes an acute atrophy of the bone similar in some respects to Sudeck's atrophy in that it develops after injury to the nerve by an operation, irritation, or trauma Other important factors in this regard are (*a*) the presence of associated extreme pain on motion and tenderness on pressure, (*b*) severe muscle spasm, (*c*) mottled rarefaction of the affected bone on the roentgenogram, (*d*) in the acute stages the periosteum may be separated from the bone, and (*e*) the condition is self-limited and heals spontaneously after an interval of three months to two years (3) Other less generally accepted theories are that the condition is due to extension by continuity of infection from the space of Retzius or other prevesical spaces, that it is due to extension of lymphatic spread of a low grade pelvic cellulitis, or is caused by injury by the needle utilized in the prostatectomy

Therapy Prevention is extremely important and includes improvement in operative technics avoidance of pre operative trauma by in dwelling catheters and unnecessary instrumentation It is essential to avoid tight packing of the prostatic cavity, prevent septic spillage and maintain adequate drainage Excellent results have been reported, particularly with reference to relief of pain with deep roentgen therapy In many cases irradiation has resulted in prompt improvement even after several months of immobilization in a plaster cast Soon after roentgen therapy the patient is able to walk and assume his normal activities Vitamin B complex has been administered with success in many instances Diathermy has also been utilized In some cases a combination of roentgen therapy and diathermy is most successful Rest and immobilization drugs to relieve spasm and atropine have proven effective in some instances Chemotherapy with antibiotics and sulfa drugs may also be beneficial Sedation is indicated to control pain Surgical procedures are not recommended except in the presence of a retropubic or pelvic abscess Roentgen therapy has proven most successful with small, divided doses Seventy-five to 100 r are administered every two or three weeks with 200 kv and 1 mm cu filtration until the acute symptoms have subsided and at longer intervals thereafter Hospitalization is advisable during the first treatments Auxiliary methods of treatment and plaster casts are not necessary Roentgen therapy is discontinued after subsidence of the symptoms *It is not necessary to continue the therapy until normal* bone architecture has been restored The aim is to alleviate the severe pain and shorten the duration of the disease The pain is relieved within

formation In adults the initial periostitis is succeeded by an extensive area of rarefaction The cortex is absorbed and the cancellous bone becomes coarse in appearance and expanded The cortex is eroded and there is sclerosis of the trabeculae in the adjacent portion of the shaft The cortical irregularity and sclerosis increase and the cancellous portion of the bone develops irregular areas of rarefaction which present a honeycomb appearance In the advanced stages, pathologic fractures are common The bone destruction may become so extensive that fragments of bone are extruded into the adjacent soft tissues The remaining portion of the shaft becomes widened and sclerotic The medullary cavity may be completely obliterated even in the absence of sequestrum, involucrum, or fistula formation Differentiation from syphilis, enchondroma, malignant tumor, sarcoidosis, coccidioidomycosis, leprosy, and yaws must be made Biopsy is the most reliable aid in diagnosis

Tuberculosis Involving the Pubis and Ischium

Tuberculosis of the pubis and ischium occurs in children in the prepubertal period The disease is most probably secondary to another focus in the body The incidence is highest in the age group from eight to eleven years although it may occur at any age The disease is more common in males than in females The roentgen manifestations comprise small cystic zones of increased radiance The margins of the areas of radiance show bone reaction typical of a granuloma particularly in the case of small and localized lesions In the more advanced forms of the disease there is vacuolization and sequestrum formation with preservation of the cortex Extensive and complete destruction of the bone occurs when the disease becomes far advanced A periosteal reaction may ensue with new bone encasing the affected area The more virulent the lesion, the greater the degree of decalcification and conversely the more chronic the disease the greater the increase in density and the more extensive the new bone formation Marked bone atrophy occurs Lesions of the symphysis pubis are characterized by marginal bone destruction with sequestrum formations There may be extension to the horizontal and descending rami of the pubis The radiant areas are surrounded by localized zones of increased density or atrophy depending on the stage and virulence of the disease In differential diagnosis it is necessary to include osteomyelitis osteochondritis, eosinophilic granuloma, lipoidosis, solitary myeloma and sarcoma

Tuberculosis of the Greater Trochanter of the Femur

Tuberculosis of the greater trochanter and its bursa is relatively uncommon The disease may occur at any age but symptoms usually begin during adolescence or early adult life The condition appears to be more common in males Occupation is of no importance in the etiology A history of trauma may be present but this is not of definite significance In practically all cases there is tuberculosis in other parts of the body A discharging sinus in the region of the hip is commonly present This may appear spontaneously or persist after operation In some cases the disease is limited to the bursa of the trochanter the bone itself not being involved

Roentgen Manifestations The lesion in the greater trochanter may be minimal and involve only the outermost aspect For this reason, the

tendency to spontaneous regression and skin lesions The condition is frequently confused with sarcoidosis The disease in adults differs in many essential respects from that in children, the axial skeleton showing varying degrees of involvement with no lesions in the metacarpals or the phalanges While the condition in many cases appears to be limited to one bone or adjacent bones there is frequently widely disseminated involvement with lesions in the skull, spine, ribs, scapula, ilium and other bones The process appears in many locations simultaneously or within a relatively short period of time Paravertebral abscesses may occur but are the exception rather than the rule

Tuberculous Dactylitis

Tuberculous dactylitis or spina ventosa is usually a disease of infancy and childhood and is rare in adults However, it does occur in Negroes

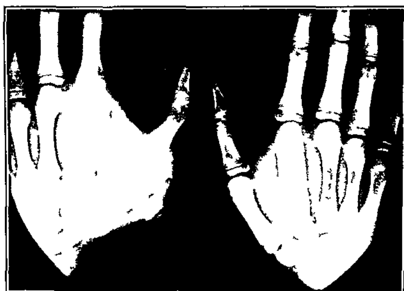


FIG 179 Tuberculosis of the Bones of the Hands There are extensive destructive changes with multiple cystic formations involving the right second metacarpal and the left first and second metacarpals The disease does not involve the joints The soft tissues in the affected areas are markedly increased in density and thickness

Chinese and others The disease is more common in childhood due to the vascularity of the short tubular bones at this period of life An important factor is the marked regenerative power of the periosteum in children the periosteum being less active in adult life The earliest roentgen manifestation is elevation of the periosteum manifested by a linear area of density surrounding the diaphysis Subsequently there is expansion and irregularity of outline of the cortex Soft tissue swelling ensues and becomes more pronounced as the disease advances In children the periosteal reaction is intense and the cortical expansion is so extensive that the normal shaft is completely encased within the expanded periosteal sheath of new bone The shaft undergoes destruction progressively and separates as a sequestrum The sequestrum shows progressive fragmentation and absorption with the formation of cyst like cavities or separates with necrosis of the new periosteal layer and fistula

heal rapidly with specific treatment Actinomycosis may produce closely similar manifestations and must always be considered in differential diagnosis Sarcoma of the sternum may simulate tuberculosis and a biopsy is necessary to establish the diagnosis Other tumors such as cysts, lipomas, and fibromas must also be ruled out in the same manner

Tuberculosis of the Ribs

Tuberculosis of the ribs is relatively easy to diagnose in the presence of other tuberculous lesions Unless local swelling is present the condition may easily be overlooked particularly if only one rib is affected A draining sinus in the region of a rib is an important aid in diagnosis There may be internal abscesses and destruction of the rib without external abscess formation Pathologic fractures may occur In many instances there is associated tuberculous mediastinal lymphadenitis The condition is more common in males than in females and is more frequently seen in adults From the roentgen point of view there are three types of lesions (1) an expanding cyst-like lesion entirely within the ribs, (2) destruction of the lower margin of the rib, and (3) complete destruction as in metastatic malignancy There is usually no evidence of new bone formation the lesion being characterized by extensive destructive changes which continue to progress slowly over a long period

Tuberculosis of the Skull

Tuberculosis of the bones of the skull is an uncommon but not rare disease The process usually begins in the cancellous bone in the diploe The incidence is estimated as about 0 2 per cent of all cases of bone tuberculosis It usually occurs in children being most common in childhood It is almost always secondary to tuberculosis elsewhere in the body practically all cases being preceded by pulmonary tuberculosis or tuberculous infections of other bones or the lymphatic glands Trauma appears to be an important contributing factor and may account for the greater incidence in boys and young children The infection reaches the skull through the blood stream usually from a distant focus This explains why the lesion in most instances appears first in the vascular cancellous diploe of the calvaria The frontal and parietal bones are most often affected The disease may occur as a single, circumscribed focus or in a diffuse progressive form Clinically there may be local swelling of the scalp headaches and tenderness The mass is firm and closely simulates tumor or cephalhematoma Fistulas may form and are apt to discharge for long periods

Roentgen Findings The lesion occurs as a circumscribed area of bone destruction punched out in character The bone and periosteum surrounding the perforation appear normal to the edge of the defect there being no osteophytes or increased density A narrow zone of rarefying osteitis may be observed at the margins of the lesion The defect may be round oval or quadrilateral The diameter varies from a few millimeters to several centimeters Sequestra of varying size may occur In the progressively infiltrating type multiple areas may be found in wide distribution and in progressive fashion The sutures offer no barrier to the spread of the process Cold abscesses and fistulas form in the soft parts as with tuberculosis of other bones

diagnosis is frequently not established because of the dark film which is obtained in the routine study of the pelvis and hip and the resultant failure to visualize the detail of the trochanter and the adjacent soft tissues. It is essential to examine the roentgenogram with a concentrated, intense light. Many cases are best demonstrated by the use of soft tissue techniques. In this manner, small calcium deposits in the trochanteric bursa and beginning erosion of the cortex can be best demonstrated. The principal roentgen manifestations comprise localized destruction of the cortex of the greater trochanter, particularly along the superolateral aspect and diminished density of the adjacent bone due to atrophy. Reactive bone or involucrum is not present unless there has been a previous operation with disturbance of the periosteum or secondary infection associated with a draining sinus. Associated bursitis is manifested by the presence of amorphous calcific depositions and swelling of the soft tissues overlying the trochanter. In rare instances areas of bony density may be present in the soft tissues in association with irregularity of the margins of the outer portion of the trochanter. Sinuses may develop and on injection with an opaque medium are found to extend to the bone.

The condition can usually be differentiated from acute infection and tumor without difficulty. Simple bursitis may involve the trochanteric bursa with symptoms and roentgen findings similar to those in subdeltoid bursitis. The presence of non-specific bursitis in the region of the trochanter is not uncommon. Calcification may occur in this bursa as in other bursas. However, this type of trochanteric bursitis responds to conservative treatment such as roentgen therapy and physiotherapy. In the absence of a sinus tract or destruction of the cortex, the diagnosis of tuberculosis cannot be established.

Tuberculosis of the Sternum

Tuberculosis of the sternum is rare. The disease affects both sexes equally. The age incidence varies from early infancy to late in life with no preponderance of any age group. *There is usually no history of* trauma. The diagnosis must be suspected when the patient with a tuberculous focus elsewhere in the body develops a swelling of the anterior chest wall. In practically all cases the disease occurs in association with other tuberculous lesions. A painless swelling in the region of the sternum is the usual presenting symptom. A discharging sinus is an important manifestation. About a third of the cases develop multiple sinuses. Any portion of the sternum or its adjacent structures may be affected. Study of the material discharged by the sinuses may be the only means of establishing the diagnosis.

Roentgen Manifestations. The roentgen manifestations comprise areas of destruction with irregularity of outline. The changes may be localized or extensive. A segment or the entire sternum may be involved. As the sternum is difficult to show in routine roentgenograms the radiologist must be aware of the fact that this disease is under consideration and utilize special lateral and oblique projections. Laminography is of particular value and may reveal the lesions when other methods fail. The condition must be differentiated from tuberculosis of the chest wall, breast, ribs and costal cartilages. The injection of radiopaque substance into the sinus is an important aid in diagnosis. Syphilis can be excluded by the Wassermann test and the fact that luetic lesions usually

or at least are never diagnosed The condition is frequently confused with tuberculosis, which is an additional factor adding to the difficulty of establishing the incidence of the disease Sarcoidosis may appear at any age Most cases occur in the period from fifteen to fifty years, the highest incidence being in the *third and fourth decades* It is rare under the age of ten and after the age of sixty There is an apparent predominance in the female, the ratio being 2 to 1 or more While the disease is relatively common in the white race, especially in the Nordic group, the incidence in this country is definitely higher among the Negroes Reports of unusually high incidence in several members of the same family have been recorded The number of cases with bone involvement is not known Osseous disease is more common in European countries than in the United States Negroes appear to present bone lesions more frequently than the

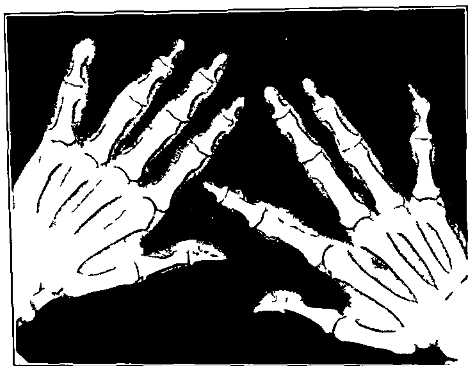


FIG 180 Sarcoidosis The terminal phalanges are markedly thinned and eroded and show multiple cystic changes There is little or no osteoporosis The distal phalanx of the left little finger is practically completely destroyed The patient had sarcoidosis of many years duration

white race Holt indicates that about 15 per cent of patients with sarcoidosis have involvement of the bones

Clinical Manifestations The manifestations of sarcoidosis are protean and lesions have been described in *every portion of the body except the hair and nails* There is an endless variety of clinical forms with transitions from one form to the other Lesions of particular interest to the roentgenologist occur in the lungs the skeletal system the heart the stomach the intestinal tract and the brain The first description of osseous sarcoidosis was made by Kienbock in 1902 about three years after Boeck s description of the manifestations in the skin and the lymph nodes The correlation of the skin and bone lesions was first made by Kreibich in 1904 who described a patient with lupus pernio and destructive changes in the phalanges Schaumann and Jungling established that the bony defects were due to a granulomatous

Differential Diagnosis The principal conditions which must be considered in differential diagnosis are syphilis, osteomyelitis and malignant neoplasms Syphilis is more common in adults than is tuberculosis The roentgen changes in syphilis are apt to consist of multiple zones of bone destruction which vary in size are separated from each other by areas which are normal or sclerotic, and are not sharply circumscribed The skull may be thickened, uneven, and have a moth eaten appearance Osteomyelitis is more acute and is accompanied by fever, pain, and edema of the soft parts the bone changes, however, may be quite similar to those in tuberculosis Osteosarcoma develops slowly, is more irregular and is less sharply circumscribed, spicule formations may be present on the roentgenogram In perforating malignant neoplasm, the roentgen changes are those of a purely destructive lesion with irregular, poorly outlined borders and there is at times a soft tissue mass which is irregular, firm, and nonpulsatile

SARCOIDOSIS

In 1899 Boeck published a report of the disease which has since been associated with his name He assumed that the lesions in his patients were sarcoma-like and believed them to be of tuberculous origin The condition is now known quite generally under the name of sarcoidosis, Boeck's sarcoid, benign lymphogranulomatosis and Besnier-Boeck-Schaumann's disease Although Boeck was the first to describe the histologic characteristics of the skin lesions, Hutchinson in 1875 had published a clinical report of such cases Besnier in 1889 and Tenneson in 1892 had also described the skin manifestations of the disease Later it was found that other parts of the body could be affected and in 1914 Schaumann postulated that it was a generalized disease which affected primarily the lymphohematopoietic apparatus It is now known that such conditions as uveoparotid fever and some cases of Mikulicz syndrome are manifestations of the same entity The disease has been known under many names, the chief of which are Hutchinson's disease Besnier's lupus pernio Boeck s multiple benign sarcoid Heerfordt's uveoparotid fever, Schaumann s lymphogranuloma benignum, and the Mikulicz syndrome Many are of the opinion that sarcoidosis is a peculiar manifestation of tuberculosis and there is extensive data which appears to support this theory However sarcoid like lesions can be produced locally by the injection of other organisms than the tubercle bacilli and also by numerous inert substances such as grass and silica Some authorities believe that sarcoidosis is a disease of diverse etiology or that it is due to an unrecognized bacillus fungus, or filterable virus Much has been written on the etiology of sarcoidosis There is no agreement as to the underlying causative factor

Incidence While formerly considered a rare and unusual condition of interest only to the dermatologist, it is now known that the disease is widely disseminated and involves the chest and other organs Large series of cases have been recorded in Scandinavia Australia, Japan, Latin America United States Canada and other parts of the world The distribution seems to be world wide with an apparent predilection for the cooler countries Roentgen study of over a half million recruits for the Swiss army showed an apparent incidence of 13 cases per 1000 Since the disease is relatively benign it is apparent that many cases are overlooked

or heart shaped They are solitary or multiple and occur in any portion of the bone The small cysts may fuse to form large, sharply circumscribed defects The bone adjacent to the cystic areas is entirely normal in appearance In some instances there is complete destruction of the trabeculae of the cancellous bone with thinning and expansion of the cortex producing an appearance very similar to that of the so called spina ventosa in caseating tuberculosis In the early stages there is diffuse osteoporosis This is followed by a non specific deossification which is supplanted by a reticular pattern of bone destruction and constitutes one of the most constant and characteristic findings of the disease The reticular pattern may be described as a lace work grill work, or lattice-work The medullary and perivascular lesions may occur simultaneously and in varying combination, hence transition forms are of common occurrence The most important and easily recognized diagnostic combination of roentgen signs is the diffuse reticular pattern plus one or multiple cyst-like areas in the region of the nutrient foramen Pathologic fractures are common and indicate marked extension of the disease with advanced destruction of bone Mutilating deformities may result and necessitate amputation

The changes may undergo partial or complete regression spontaneously, particularly in the case of diffuse involvement As a rule the bone is not restored to an entirely normal appearance some residual alteration persisting and resulting in a permanent deformity The cyst-like vacuole in the end of the bone is apparently the most chronic and persistent lesion The diffuse lace like lesion in the phalanges may resolve into vacuoles or pseudocysts and persist for many years The periosteum is not involved and there is usually no destruction of the adjacent joints or subperiosteal new bone formation as with inflammatory disease The absence of periosteal thickening is extremely important in establishing the diagnosis from the roentgen point of view When periostitis is present the lesion most probably is not sarcoidosis There are no bone sequestra or draining sinuses in the disease Despite the extensive involvement of the bone, there is normal or practically normal function of the joints, even when severe mutilating deformities are present The osseous lesions are painless and in consequence there is no disuse osteoporosis of the affected extremities The cartilage of the joints remains intact Polyarticular sarcoidosis has been described in the younger age groups but this is extremely unusual In children the epiphyses are apt to be involved

Differential Diagnosis Many lesions must be considered in differential diagnosis Hyperparathyroidism and polyostotic fibrous dysplasia may present changes in the phalanges which closely simulate the advanced, diffuse type of sarcoidosis When the digits are involved in the fibrocystic diseases the other long bones of the skeletal system show even more extensive involvement Multiple enchondromata usually expand the cortex of the involved bone to a much greater degree than is the case in sarcoidosis In gout there are tophi with sharply defined smooth cyst like margins Similar changes occur in rheumatoid arthritis and osteoarthritis Sarcoidosis practically never involves the joints and this is important in establishing the diagnosis The systemic fungus diseases sporotrichosis blastomycosis torulosis and coccidioidomycosis may produce destructive lesions in the phalanges in association with bizarre pulmonary lesions The bone changes in leprosy may closely simulate those of sarcoidosis Yaws must be included in the differential diagnosis In many instances the clinical course of the disease must be observed

process identical microscopically with the changes described by Boeck The bone lesions of sarcoidosis have a predilection for the small bones of the hands and feet The middle and distal phalanges are the most common sites, the proximal phalanges, the metacarpals and the metatarsals being involved occasionally and other bones only rarely Sarcoidosis has been reported in practically every bone of the body although only in the bones of the hands and feet are the changes of diagnostic significance The bones of the feet are usually more extensively involved and show more marked changes than the hands, although this is not universally the case It is advisable to make roentgen study of both the hands and the feet periodically in order to detect the changes in their early stages An important point suggested by Holt and Owens is that detailed roentgenograms of the nasal bones should be included as part of the skeletal survey in patients who present dermatologic manifestations of sarcoidosis as involvement of the bridge of the nose is common, direct extension of the process not infrequently producing painless, non tender destruction of the nasal bones Failure to find the characteristic defects in the bones does not exclude the diagnosis Pronounced fusiform swelling of the fingers may be present in the absence of roentgen manifestations of the disease The incidence of bone lesions appears to be directly proportional to the incidence of skin involvement This applies whether the skin changes are located on the face, back fingers, or toes There is no appreciable difference in the age and sex of patients with osseous lesions of sarcoidosis as compared with manifestations of the disease in other organs

Pathology The histopathologic changes in sarcoidosis of the bone are the same as in other tissues There is a granulomatous reaction consisting of a basic tubercle composed of epithelioid cells, giant cells, and lymphocytes There is no evidence of caseation necrosis, which constitutes the significant factor in the differentiation from tuberculosis The giant cells are larger than in tuberculosis there being 25 to 30 nuclei uniformly distributed in each cell rather than elliptically arranged as in tuberculosis The histologic diagnosis of sarcoidosis is not accurate in many instances It is generally accepted that microscopic examination of an excised lesion may indicate the presence of the disease but definite diagnosis must be based on the sum total of the manifestations in the individual case

Roentgen Manifestations The skeletal lesions in many instances are limited to the small bones of the hands and feet This is particularly significant because the disease is bizarre in its clinical manifestations and every aid in diagnosis is extremely important The bone changes are characteristic and in many instances permit of the establishment of the diagnosis with definiteness The medullary cavity is the primary site of the epithelioid tubercles of sarcoidosis and the lesions are frequently more extensive than the radiologic examination indicates Sarcoidosis in the marrow is usually associated with changes in the adjacent bone The changes may occur in one or several patterns In the early stages there is a stippled pattern with tiny dots of diminished density projected against the background of bone which is normal or nearly normal in density The rarefaction is more marked at the distal ends of the proximal and middle phalanges and the proximal end of the distal phalanges There is enlargement of the lacunar spaces with coalescence which results in the formation of cavitations of widely varying size and shape These are termed the punched out lesions and may be rounded ovoid, pear shaped

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before the diagnosis can be established Tuberculous dactylitis of the so called spina ventosa type occurs in children and involves the metacarpals and the metatarsals more extensively than the phalanges There is usually periosteal thickening and this in combination with the clinical findings establishes the diagnosis The lesions of cystic tuberculous osteitis may closely simulate the manifestations of sarcoidosis although the clinical aspects of the disease are different Not infrequently there are small, isolated rounded areas of increased radiance in the bones of the hand in entirely normal individuals These vacuoles are most common in the heads of the metacarpals and represent cartilaginous or fibrous tissue replacement of bone resulting from a minor defect in normal ossification and are not of pathologic significance

OSTEOMYELITIS DUE TO BACILLUS PARATYPHOSUS A

A rare cause of osteomyelitis is the Bacillus paratyphosus A The lesion is osteolytic in nature with massive destruction of the affected bone The picture may closely simulate osteogenic sarcoma In some instances there is extensive new bone formation and sequestration in association with the osteomyelitis It is essential to differentiate the condition from atypical bone infections and osteogenic malignant tumors Spondylitis caused by the bacilli of the typhoid paratyphoid group is rare The disease as a rule begins during the period of convalescence from the primary infection but may occur earlier or later Pain tenderness and limitation of motion are the chief clinical manifestations This is followed by the development of rigidity and changes in spinal curvature The roentgen manifestations may not appear until the late stages of the disease There is absorption of the intervertebral disc with slight destructive changes in the body of the vertebra Bone proliferation with deposition of new bone along the lateral spinal ligaments develops and results in bony ankylosis of the adjacent vertebral bodies Widening of the paravertebral soft tissue shadows indicating abscess formation is rare but may occur

SYPHILIS OF BONE

Syphilis of bone is divided into two types (1) congenital syphilis which may manifest itself at birth during childhood or not until puberty or adult life and (2) acquired syphilis The acquired form of the disease usually does not present skeletal manifestations until many years after the primary infection has taken place Since the roentgen changes in congenital and acquired lues are widely different it is necessary to describe each form of the disease separately

A **Congenital Syphilis** Early cases which develop during intrauterine life and are demonstrable in the neonatal period are characterized by a zone of increased density with irregularity of outline in the region of the junction of the epiphysis and diaphysis due to calcific depositions in the epiphyseal cartilage There is a narrow band of increased radiance at the end of the diaphysis resulting from epiphyseal separation Slight periosteal thickening may be present The changes involve multiple bones At a later stage there are irregular areas of bone destruction or zones of rarefaction in the metaphysis adjacent to the

epiphyseal line The epiphyses are not affected Periostitis is present along the shafts of the long bones with elevation and proliferation of the periosteum Bone atrophy is absent and pathologic fractures seldom occur The central portions of the long bones show increased density Ossifying periostitis ensues during the first year of life with double contours due to new bone formation along the surfaces of the shafts or ends of the long bones After adequate treatment, the lesions disappear completely and the bones become normal in appearance

Congenital tertiary lues is essentially similar to acquired lues in the adult and comprises involvement of the shafts of the long bones Extensive new bone formation and areas of destruction are common In some cases periosteal and endosteal bone proliferation are the sole changes



FIG 181 Congenital Lues There is involvement of the bones of the extremities with periosteal proliferation and areas of rarefaction at the ends of the long bones

In other instances there are areas of bone destruction with sharply defined borders scattered in the cortex and complete absence of bone within the affected area due to gummas Untreated congenital syphilis results in thickening and increased density while untreated acquired lues results in thickening, increased density and curvature of the long bones The tibia and the bones of the forearm are most frequently affected The bowing is usually anteriorly and the changes are most marked along the convexity or anterior aspect of the bone The "sabre" deformity of the tibia is the typical manifestation of this condition The prominence of the trabeculae and cortical thickening may closely simulate Paget s disease and differential diagnosis is difficult In Paget s disease the trabeculae are more wavy and the bone less dense than in lues Skeletal

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superimposition of newly formed bone A characteristic example is the deposition of bone at the anterior surface and crest of the tibia imparting a curved appearance with an anterior convexity There is a striking difference between the gumma and diffuse syphilitic inflammation of bone In the latter, there is superficial invasion of the cortical bone, and only the outer layers are destroyed In the presence of a gumma, there is extensive loss of bone substance

Pathologic fractures are not infrequent, being more common in the neurogenic forms of syphilis It is to be expected that fractures would be rare in view of the fact that the usual bone reaction is that of proliferation

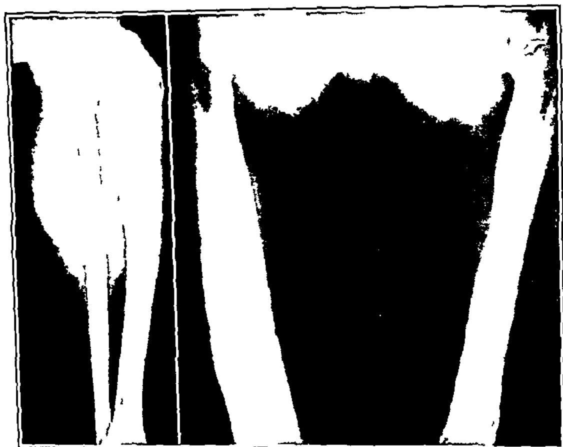


Fig 182 Syphilis The right lower leg and both femurs There is marked cortical thickening bowing and narrowing of the medullary cavities The changes are more advanced on the right

rather than destruction However in many instances there is extensive osteolysis and the pathologic fractures when they do occur usually involve the areas of bone destruction There is widespread dissemination of spirochetes through the body tissues within three to four hours after the initial infection Despite the fact that the bone periosteum and marrow cavity are invaded by myriads of spirochetes skeletal involvement is comparatively rare

Syphilis of the Skull

Syphilis of the skull is becoming increasingly uncommon owing to modern methods of prophylaxis and early therapy It is seen with sufficient frequency however to require a thorough knowledge of its manifestations It must always be borne in mind and considered in the differential

hyperostoses may occur in congenital syphilis and when the process is periosteal, the changes simulate those of benign bone tumors With endosteal bone changes, the medulla may be partially or completely obliterated In the presence of involvement of the bones of the skull and face marked deformities which do not respond to antiluetic therapy ensue and may require surgical removal

Spirochetes have been demonstrated in the periosteum, marrow and cartilage after the fifth intrauterine month in the syphilitic fetus The osseous lesions may be manifest four to six weeks later although in most cases they are not demonstrable until after birth The early lesions rarely appear after the age of six months The long bones are most frequently involved, the metacarpals phalanges and ribs less commonly Involvement of the skull, jaw and spine is exceptional in the infant The lesions tend to affect all the long bones In both acquired and congenital syphilis, periostitis, gummatous changes hyperostosis and osteitis may be present Osteitis or hyperostosis usually occurs in the late forms of the congenital type of syphilis It must be stressed that the bone lesions of congenital syphilis may be delayed in appearing until puberty or even as late as the twenty fifth to thirtieth year of life From the age of six months to seven years, few syphilitic osseous lesions are seen From seven to fourteen years the tardive lesions appear Osteochondritis is rare Rarefying osteomyelitis is infrequent Condensing osteomyelitis accompanied by periostitis is the usual picture and most frequently involves the tibia

B **Acquired Lues** Any bone may be involved The lesions in the tertiary stage develop eighteen months to many years after the onset of the disease The skull and long bones are most frequently affected The process is characterized primarily by bone proliferation and in the long bones there is irregular fusiform, dense thickening of the cortex of the bone As a rule, there are no changes in the medullary cavity There may be osteitis periostitis osteomyelitis, arthritis and gumma formation In luetic osteitis, dense sclerotic bone is formed The entire cortex is involved The shaft is not expanded and there is no change in the adjacent soft tissues In luetic periostitis spicules of new bone project at right angles from the shaft of the bone forming a fine network of trabeculæ The tibia and ulna are most frequently involved When the phalanges are affected the process is termed dactylitis In the tibia the involvement is along the anterior surface and produces the sabre tibia Luetic osteomyelitis simulates pyogenic osteomyelitis roentgenologically Clinical evidence of active inflammation is absent There is simultaneous bone proliferation and destruction with no expansion of the shaft The lesion usually originates in the medullary portion There may be zones of apparently normal bone interspersed with the involved areas There is no reaction in the adjacent soft tissues Gummas usually occur within an area of increased bony density and present a rounded or irregular area of increased radiance The margins are irregular and poorly defined The tibia is most frequently involved In the skull the frontal bone is the commonest site the parietal and other bones being less commonly involved The syphilitic gumma may originate in the bone or extend to the bone from the adjacent soft tissues Syphilitic osteoperiostitis comprises a more diffuse lesion which originates in the periosteum The syphilitic form of osteitis particularly osteoperiostitis is characterized by extensive formation of new bone The affected bone is deformed by the

Syphilitic Spondylitis

Syphilitic spondylitis is the result of invasion of the periosteum and bone of the vertebra by the treponema pallidum. Syphilis of the vertebra should not be confused with Charcot's disease of the spine as the latter is secondary to changes in the dorsal root ganglia and columns of the spinal cord with resultant loss of sensory perception and impairment of the reparative mechanisms of the traumatic effects of daily life.

Pathology. The changes in syphilitic spondylitis are similar to those in syphilis of other bones. The disease begins in the periosteum and comprises a periostitis. Edema of the connective tissue ensues. If healing does not take place there is periosteal new bone formation and osteophytes or exostoses form. New lamellae are deposited on the Haversian canals with condensation and eburnation of the involved bone. Osteoperiostitis and osteomyelitis result in the formation of a gumma with extensive necrosis of the bone. The discs may be involved in the gummatous process and undergo destruction. A superimposed secondary infection may ensue and complicate the picture.

Since the cervical region is the most freely movable part of the spine, this may be the explanation for the relatively greater frequency of involvement of this segment of the vertebral column. Spondylitis develops in the virulent form of the disease. It may occur in congenital syphilis or during the second or third stages of acquired syphilis. The history of a primary or secondary syphilitic lesion is of little importance. Serologic tests may be negative.

Clinical Manifestations. The symptomatology is not definite and clinical diagnosis is usually impossible. The normal lordosis of the spinal column is obliterated. The affected segment of the spine is rigid. Localized pain may be more marked at night with tenderness and stiffness which are due to the associated periostitis. Compression of the cord is seldom sufficiently severe to produce compression myelitis. Co-existing lesions in other bones are common, particularly the sternum, clavicle, tibia, and radius. Sudden death may occur due to erosion into an artery.

Roentgen Manifestations. Any portion of the spinal column may be affected. There is usually destruction of the body of the vertebra, particularly its ventral aspect. There is marked sclerosis of the bone with hyperostosis and eburnation. Calcific depositions develop in the anterior and lateral ligaments and may lead to complete ankylosis. Eburnation predominates over destruction. The intervertebral space may be narrowed or remain normal in width. The vertebral body may become rounded and larger than normal in size. The normal bony structures are replaced by sclerosis or areas of destruction. The two manifestations may co-exist. In the cervical region the ligaments ossify in a uniform, smooth fashion. In the lumbar and dorsal regions large pointed spurs develop along the anterolateral borders of the end plates. The picture at this stage may closely simulate degenerative arthritis. However, osteoarthritis is usually a generalized condition involving all or large sections of the vertebra while the spur formations in syphilitic spondylitis are limited as a rule to 2 or 3 vertebrae. There is usually no paravertebral abscess. Sequestration may occur. Subluxation and compression of the vertebral body are common. In rare cases the picture may be similar to that in hemangioma with a mottled, irregular appearance and a cuneiform

diagnosis of every bizarre and unusual case, as it may mimic many other conditions

Roentgen Findings There is no single series of manifestations which may be considered pathognomonic Here, as elsewhere in the body, syphilis is the great imitator and may closely simulate many other lesions In gummatous processes, there is bone destruction with necrosis These findings are most common in the frontal and parietal bones, frequently being limited by the lambdoidal suture although the posterior portion of the skull may also be affected Both the inner and outer tables are destroyed There is extensive resorption of bone The destruction may follow the paths of the blood vessel grooves Multiple, scattered areas of rarefaction occur as in osteomyelitis with coalescence in serpiginous

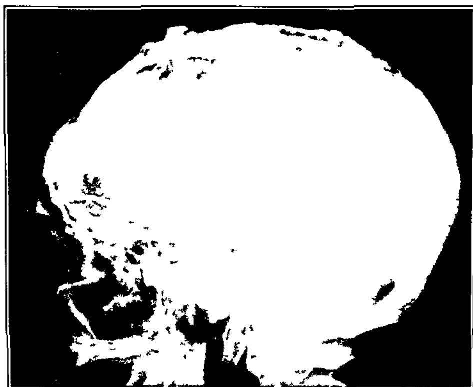


Fig 183 Syphilis of the Skull There is extensive bone destruction in the frontal and parietal regions The changes involve both the inner and outer tables and are characterized by irregular areas of increased radiance with segments of normal bone intervening The condition had been present for over ten years with slow progression despite therapy

forms Parrot s nodes with small rounded sharply defined areas of bony density projecting from the outer tables of the skull may occur in adults Sequestra form and may discharge to the surface New-bone formation gives periosteal thickenings and osteophytes Scattered patches of increased density due to calcifications along the periosteal layers and condensing osteitis may occur Thickening of the tables widened diploic spaces and scattered areas of increased density throughout the skull may form in the calvaria and extend into the soft tissues as in meningioma and sickle cell anemia Areas of destruction with patches of increased density may give an appearance similar to that in myeloma and metastatic carcinoma The changes may be so varied as to imitate almost any other lesion seen in the skull A thorough knowledge of the history and clinical findings is essential for accurate diagnosis

ACTINOMYCOSIS

Actinomycotic infection is due to a microaerophilic or ray fungus The fungus may lie latent in the crevices of carious teeth or in the tonsils and is swallowed or aspirated into the lungs It does not penetrate epithelium without the aid of local trauma or an underlying pathological process However, once having obtained a foothold, it can grow and spread widely in the connective tissue causing a chronic, suppurative lesion The disease usually originates in the soft tissues and involves the bone secondarily, resulting in a localized osteomyelitis Bone tissue reacts to actinomycotic infection in a manner which is characteristic and differs in certain respects from other forms of bone infection In the region of the focus of infection, there is absorption of bone In the area adjacent to the necrotic zone the cells are stimulated to form new bone and this may attain a



FIG 184 Actinomycosis Sagittal View of the Lumbosacral Spine There is extensive destruction of the lamina and the pedicle of the fourth and fifth lumbar vertebræ on the right and of the right fifth transverse process The intervertebral space between the fourth and fifth lumbar vertebræ is narrowed and irregular (black arrow) The changes are due to involvement of the spine by the actinomycotic process (Ritvo *Chest X Ray Diagnosis*)

marked degree of density The combination of caries and sclerosis in various proportions produces protean pictures Sequestrum formation is common The bone may become very thick and be traversed by a series of channels with sinuses in which the fungus is present Less commonly, rarefaction is the sole manifestation or predominates and large cavities develop As a rule the bone is involved by direct spread from foci in the neighboring soft tissue The spinal column lies in the path of extension of several common foci and may be involved from the neck, the mediastinum the retroperitoneal tissues after perforation of the appendix and other sources The jaw is a frequent site of the disease Actinomycosis of the spine is becoming more frequent Only a relatively small number of cases of osseous actinomycosis have been recorded in the literature This is due to the fact that many cases are not recognized during life, the diagnosis being established only at post mortem examination In many instances the disease is diagnosed incorrectly

shape of one portion of the vertebra The eburnation or sclerosis may regress after antiluetic therapy

Differential Diagnosis The differential diagnosis of congenital syphilis is concerned principally with rickets, scurvy, traumatic lesions and pyogenic infections Congenital lues is usually manifested about one month after birth with normal development prior to that time Neither rickets nor scurvy develops so early in life Syphilis is characterized by prompt improvement subsequent to antisyphilitic therapy and as a rule the bones return to an entirely normal appearance after adequate treatment In acquired syphilis, Paget's disease, tuberculosis osteoblastic metastatic carcinoma, osteogenic sarcoma, Ewing's tumor, pyogenic osteomyelitis, sclerosing osteomyelitis of the Garre type, Brodie s abscess, lymphoblastoma and many other conditions may produce closely similar changes In the early stages, tuberculosis tends to be progressive with predominant bone destruction while new bone production occurs later Lymphoblastoma and leukemia produce bone destruction, bone formation or a combination of both The extraosseous manifestations of the disease establish the diagnosis Primary sarcoma is difficult to differentiate However, simultaneous involvement of other bones is rare and the condition usually shows rapid progression in contradistinction to the relatively slow advance in lues Careful evaluation of the clinical findings and serial roentgen studies at intervals of a few days or weeks are frequently necessary to establish the diagnosis with definiteness As with other unusual lesions the diagnosis will be made only if the condition is borne in mind It is necessary to correlate the history, physical findings serology, roentgen studies and other laboratory tests The presence of active syphilis is significant Improvement with antisyphilitic therapy is not diagnostic because certain types of sarcoma appear to improve after antiluetic therapy and iodides are of benefit in actinomycosis

YAWS

Yaws is a tropical disease which has many features in common with syphilis, the bacteriologic and clinical aspects being very similar in the two diseases The bony manifestations occur in the tertiary stage In many instances there is a history of florid secondary lesions during childhood The most common sites of bone involvement are the tibia the inner end of the clavicle and the humerus The hands and feet may also be involved and skull lesions are not uncommon A periosteal reaction may develop within a few months after infection has taken place The periosteal new bone is formed in layers parallel to the shaft of the bone Subsequently there is bone erosion The areas of erosion are usually multiple vary in size from 2 mm to 3 cm in diameter and present sharply defined margins In the skull the lesions are cystic in character and frequently are surrounded by a narrow band of dense sclerotic bone Joint involvement is not common The presence of localized areas of erosion with margins of sclerotic bone and nearby periosteal new bone formation result in appearances very similar to those of syphilis In some patients soft tissue swellings are present in the affected area and the changes in this instance closely resemble sarcoma The joints may be affected with extensive destruction of the articular cartilages

loss of weight, irregular fever, secondary anemia and a moderate increase in the white cell count in the blood as in active infection. In many instances there are sinuses which extend to the external surface of the body due to operative procedures or spontaneous opening of an abscess. The diagnosis is established by the demonstration of the actinomycotic organism in the fluid obtained from the affected part. The radiographic appearances are usually not diagnostic. Therefore, it is essential that all abscesses which drain in the neck, back, or loin be studied carefully for actinomyces.

Differential Diagnosis. The principal diseases which must be considered in the differential diagnosis include chronic osteomyelitis, tabes dorsalis, tuberculosis and primary carcinoma of the lung. Other conditions which must also be included are metastatic carcinoma, myeloma, and osteitis deformans. The diagnosis of tuberculosis is frequently established erroneously in actinomycosis particularly in lesions involving the spine. In actinomycosis of the spine, the transverse processes and heads of the neighboring ribs are frequently destroyed while collapse of the vertebral body and kyphosis are uncommon. An important factor also is that the rarefaction of bone is accompanied by sclerosis. In both actinomycosis and tuberculosis large abscesses may form. The tuberculous abscess tends to extend outward from the vertebra and follow definite pathways along the psoas muscles or the ribs. The actinomycotic abscess progresses from adjacent tissues to attack the bone and drains internally or reaches the surface of the body directly. Pyogenic osteomyelitis is acute in onset and is often associated with sequestra. Rarefaction of bone may occur in osteitis deformans, metastatic neoplasm and osteitis fibrosa cystica. In these conditions there is no fever or other manifestations of chronic inflammation.

Therapy and Prognosis. Until recently the outlook was hopeless except in cases with localized, superficial lesions of the bone. Treatment was unavailing and death resulted after a period varying from a few months to two years. Drainage of abscesses, x-ray therapy and the administration of iodine compounds were purely palliative. With the use of the newer drugs the prognosis has been greatly improved and with drainage of abscesses and the maintenance of nutrition a favorable outcome may be foreseen. The essential treatment is the administration of the antibiotics, preferably penicillin, and the sulphonamides. The fungus may remain latent and recurrences are common. Treatment should be continued long after the patient appears to be clinically well. It may be interrupted when the radiographic appearances show that new bone has formed in the affected area, indicating that the process has been arrested and has undergone healing.

BLASTOMYCOSIS AND COCCIDIOIDOMYCOSIS

The mycotic infections of bone, while relatively rare, must be considered in all cases of chronic infection in which the manifestations are atypical and the diagnosis cannot be established readily. Of the group of fungus infections which may affect the bone, among the most common are blastomycosis and coccidioidomycosis. The manifestations of these pathogens will be considered together as they are similar and cannot be differentiated from each other by roentgen methods.

The changes in the bones vary in extent and intensity The disease may be limited to one bone but more commonly affects several bones In the case of the spine large segments of the vertebral column are involved as the process spreads along the anterior ligaments The intervertebral discs are frequently preserved in the early stages, as the disease advances, the discs are involved It differs from tuberculosis in that the lesion spreads to the adjacent pedicles, the transverse processes, and the heads of the neighboring ribs Rarefaction and sclerosis are present in varying proportions In the early stages or in mild cases there is absorption of the superficial portion of the vertebral body and slight deep rarefaction with little or no sclerosis In the presence of advanced involvement, either by continuous extension or vascular embolism, a characteristic picture results The bone is riddled by a network of suppurating channels which are bounded by areas of increased bone density resulting in an appearance of a honeycomb or lattice work on the radiograph During the progression of the lesion, rarefaction is more prominent As improvement takes place, sclerosis dominates the picture The process may be characterized by the formation of osteophytic outgrowths with irregularity of outline and increase in the size of the bone Collapse of the vertebral bodies may ensue and result in marked mobility of the spinal column Actinomycosis of the vertebral column is manifested in some instances by a soap bubble appearance of the spine with multiple areas of radiance interspersed with zones of normal or dense bone Paravertebral soft tissue swelling extends about and beyond the area of bone involvement and is fusiform in shape This is termed the primary or intrinsic form In the secondary, or extrinsic variety, the disease spreads to the spine from a focus in the thorax or the abdomen Dense, long spurs extend longitudinally and may bridge adjacent vertebre There may also be dense eburnated bone extending from the lateral margin of the vertebral body into the central portions of the body with replacement of the trabeculæ by a homogeneous shadow The psoas muscle shadows, renal outlines and peritoneal fat lines may be obliterated

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Pathogenesis The source of the infection is usually the soil which sends up delicate spores The spores separate from the aerial hyphae and are so light that they easily float in air The portal of entry into the human body is usually through the lungs the spores being inhaled The disease may also be contracted while performing autopsies, during the treatment of cutaneous lesions, and in the laboratory The life cycle of the spores is divided into two phases, a vegetative and a parasitic The parasitic phase occurs in the body of the infected host After inhalation of the spores of coccidioidomycosis, reproduction occurs within the tissues by rupture of the wall of the spore or microspherule with release of many endospores The endospores closely resemble cocci and give the disease its name In blastomycosis, reproduction within the tissues is by budding rather than by endosporulation This serves as an important aid in diagnosis



Fig 185 Blastomycosis of the Spine There is a localized area of destruction involving the superoanterior aspect of the body of the fourth lumbar vertebra The intervertebral space between the third and fourth lumbar vertebræ is decreased in width indicative of narrowing of the intervertebral disc

Symptoms The onset of the disease is *frequently insidious with the result* that the infection becomes widespread before the lesion is diagnosed The subacute primary respiratory phase is silent and asymptomatic Coccidioidomycosis is endemic in the Southwest, particularly in the San Joaquin Valley of Southern California and is much more widespread than blastomycosis Coccidioidomycosis pursues a fairly typical course Most of the residents of the Southwest have had a primary coccidioidodal infection and have developed an immunity to the disease An apparently benign upper respiratory infection, it has an incubation period of approximately three weeks after which an acute pneumonitis develops In most cases, this heals with fibrosis and occasionally with cavitation The relation to a fungus infection is generally not recognized and the condition is diagnosed as influenza or pneumonia About 2 to 5 per cent of these with previous respiratory symptoms develop an allergic type of reaction 2 to 10 days later with fever arthritis and muscle pains The joints are tender painful and slightly swollen but no effusion occurs The symptoms usually disappear after about a month without sequelae in the joints In the vast majority of cases nothing further ensues In a small number of patients the infection becomes systemic and a progressive secondary granuloma develops The disease affects various portions of the body and granulomatous lesions occur in the soft tissues the bones the joints, the muscles the skin and the meninges

Blastomycosis is a North American disease and is not endemic in any particular locality The onset is more insidious than in coccidioidomycosis After an interval of weeks or months following an unrecognized primary

infection, the patient develops a low grade fever with loss of weight and strength and night sweats. An ulcer of the skin or a subcutaneous abscess may be the first manifestation which suggests the presence of a fungus disease. The lesion begins as a soft, subcutaneous nodule, the overlying skin having an erythematous hue. The nodules tend to suppurate with discharge of bloody or purulent material. The abscess may heal leaving a dense, depressed scar or develop *into a chronic ulcerative lesion*. A subcutaneous abscess which forms by extension from infection of the bone usually develops discharging sinuses which persist for months or years. Blastomycosis or coccidioidomycosis should be suspected in all cases with chronic discharging sinuses from subcutaneous abscesses. There is little or no fever or leucocytosis. The disease must be differentiated from tuberculosis, syphilis, neoplasm, lung abscess, sarcoidosis, silicosis, osteomyelitis, psoas abscess, actinomycosis, histoplasmosis, sporotrichosis and moniliasis.

Roentgen Manifestations. The bone lesions of blastomycosis and coccidioidomycosis are identical and cannot be distinguished from each other. The process usually arises in the cancellous bone and is predominantly destructive with little or no periostitis, bone production, or marginal reaction. A clear cut area of complete bone destruction is the most characteristic manifestation. The bone changes are similar to those in osteomyelitis. The disease is apt to involve the bone at points of bony prominences such as the poles of the patellas, the acromion and coracoid processes, the angles of the scapula, the olecranon, the styloid process of the radius and ulna, the epicondyles of the humeri, the extremities of the clavicles, the malleoli, and the tuberosities of the tibias. There are frequently marginal solitary lesions in the ribs and sharply defined, destructive lesions of the outer table of the skull. Involvement of the spine is characterized by destructive processes involving the vertebræ and attacking indiscriminately the body, the transverse and spinous processes, and the neural arch. The disease involves the spongiosa, is predominantly destructive with little tendency to heal by bone production, and is frequently associated with pulmonary involvement. The infection in the bone is usually rapid in onset and development and is associated with little or no atrophy of the adjacent bone. The margins of the defect are irregular and poorly defined. The radiolucent area in the bone is associated with marked thinning or destruction of the cortex. The process may be solitary or involve two or more bones adjacent to each other or in different regions of the body. Faintly outlined linear calcifications may be present in the soft tissues adjacent to the affected bone(s). In many instances pulmonary involvement is not present in patients with disseminated coccidioidomycosis.

Blastomycosis of the spine closely mimics tuberculosis. The changes comprise marked narrowing of the disc spaces and collapse of one or more vertebral segments. Bone destruction is predominant and there is no proliferation for long periods of time. The disease extends from segment to segment by dissection under the spinal ligaments with the production of concavities along the anterior vertebral borders. The process may skip one or more vertebræ to involve others at a distance. Rib destruction with slow bone proliferation and subsequent sclerosis occur by direct extension or pressure erosion. Paravertebral involvement is common and is manifested by a rounded or fusiform area of soft tissue density at a single or multiple levels. The abscess may be unilateral or bilateral and in the lumbar region is manifested by bulging of the psoas muscle shadow.

Pathogenesis The source of the infection is usually the soil which sends up delicate spores The spores separate from the aerial hyphæ and are so light that they easily float in air The portal of entry into the human body is usually through the lungs, the spores being inhaled The disease may also be contracted while performing autopsies, during the treatment of cutaneous lesions, and in the laboratory The life cycle of the spores is divided into two phases, a vegetative and a parasitic The parasitic phase occurs in the body of the infected host After inhalation of the spores of coccidioidomycosis, reproduction occurs within the tissues by rupture of the wall of the spore or microspherule with release of many endo spores The endospores closely resemble cocci and give the disease its name In blastomycosis, reproduction within the tissues is by budding rather than by endosporulation This serves as an im portant aid in diagnosis



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The clinical course is slowly progressive and instances have been reported in which the disease has persisted for twenty or more years In the early stages there is pain and tenderness A hard fixed nodule develops deep in the soft tissues and the swelling gradually extends peripherally about the nodule New foci slowly develop These soften and after a few days rupture spontaneously with discharge of a fluid containing the characteristic granules After a few days of drainage the discharge ceases and the fistula heals The disease may be characterized by draining sinuses encrusted lesions and scars from old healed lesions The sinuses communicate with deep seated abscesses The swelling progresses to involve the entire foot There are few or no subjective symptoms In many instances there is pain prior to the rupture of the nodule The patient is able to walk despite the increase in the size of the foot There is usually no general systemic reaction and no enlargement of the regional lymph nodes except in the presence of secondary infection There are multiple deep seated abscesses with tortuous sinuses which communicate with each other The fascial planes offer no barrier to the infection and none of the tissues offer resistance to the disease muscle fat and bone being involved simultaneously Healing results in the formation of dense connective tissue scars with complete destruction of the tendons, bones and the anatomical landmarks On microscopic examination the organisms are found in colonies from which are sent out radiating mycelial threads A zone of leukocytic infiltration surrounds the colonies In the adjacent tissues there is a layer of granulation tissue which is heavily infiltrated with inflammatory cells Dense hyalinized connective tissue is distributed throughout the involved area

Roentgen Manifestations The roentgen manifestations comprise decalcification and atrophy of the bones of the foot and extreme swelling of the soft tissues There are multiple rounded areas of destruction in the bones of the phalanges the metatarsals and the tarsals The areas of destruction are usually in the central portion of the bone The smaller bones in some instances show expansion and cystic changes in the affected region Sinus tracts extending from the margins of the bone produce a half-moon type of defect There is absence of surrounding bone reaction or sclerosis In some areas there is complete destruction of bone Adjacent bones may show fusion particularly in the tarsal region This is believed to be due to an old productive periostitis The bones of the entire affected area show extreme decalcification and cast very faint shadows on the roentgenogram

LEPROSY

According to the specifications of the International Congress of Leprosy held at Cairo in 1938 there are two types of leprosy (1) The nerve or neural type denoted by the letter N includes all cases of benign leprosy with polyneuritic manifestations such as disorders of sensation trophic changes paralysis mutilations or macules of non leprous nature termed the leprides (2) The lepromatous type formerly termed the cutaneous is denoted by the letter L In this group are included all cases of malignant leprosy of unfavorable prognosis The lesions are found especially in the skin and other organs and as a rule show polyneuritic manifestations

Pathology The disease is a granuloma similar to tuberculosis The submiliary, miliary, or larger nodules closely resemble tubercles and may caseate The caseous material liquefies and cavities containing pus like material are produced In some instances there is suppuration from the beginning and while this may be acute, it is usually chronic and leads to abscesses and ulcers The lesions are practically always progressive with a marked tendency to dissemination through the lymph and blood streams Healing is rare but occurs occasionally Skin involvement may be manifested as nodes or papular eruptions Ulceration may ensue with sinuses leading to a deep abscess The abscesses may be limited to the subcutaneous tissues Lymph glands are involved early and extensively with caseation and suppuration The lesions in the bone are those of destructive osteomyelitis and may originate centrally, subperiosteally, or by extension from adjacent tissues

Differential Diagnosis Tuberculosis is usually confused with the mycotic lesions In tuberculosis, the joints are involved early and by predilection while in the mycotic diseases joint involvement is rare Metastatic carcinoma, myeloma, sarcoma, the various types of bone cysts, and giant cell tumor may produce similar roentgen alterations but the clinical manifestations usually permit of differentiation The diagnosis is established with definiteness by direct examination of the material obtained from the cutaneous lesions or by aspiration Microscopic examination is performed by use of a loop and a drop of saline in 10 per cent potassium hydroxide The blastomycoses occur in the preparations as single or budding spherical cells and yeast like bodies The walls of the cells are thick refractile, and form a double contoured appearance in fresh preparations The coccidioides are uniformly large, non budding, and may contain spherules The walls also are refractile The infected material may be cultured on Sabouraud's medium and after ten to forty days there develops a white cottony growth which later changes to tan or brown Animal inoculation may be utilized and permits of the establishment of the diagnosis in three weeks as typical lesions develop in the liver spleen lungs and lymphatics The prognosis is uniformly bad The mortality rate is about 90 per cent in patients who have been observed for two years or longer

MYCETOMA PEDIS

Mycetoma pedis or Madura foot was first described by Gill in 1842 The term mycetoma is used to indicate a group of infections caused by mycotic organisms and in which granules comprising the organisms are present in the tissues Madura foot occurs particularly in the tropical portions of the world and is not uncommon in India Africa Central America, South America and the Netherlands East Indies In temperate climates it is more unusual although an increasing number of cases are being reported from Canada and the United States particularly in the South The usual method of infection is from the soil after an abrasion of the foot The disease in consequence occurs most commonly in laborers particularly farmers who do not protect the feet with shoes A history of trauma and contact with soil is not present in every case The causative agent of mycetoma may be any one of a variety of species of actinomyces or true fungi The pathologic changes and the clinical course are similar despite the variability of the etiologic agent Only on the basis of laboratory

studies is it possible to differentiate the various types of causative organisms

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only when they are very far advanced or have become neural secondarily The two types are subdivided according to the degree of involvement first neural degree is designated as N1 moderate neural, N2 advanced neural, N3, first degree lepromatous, L1, moderate lepromatous, L2, and advanced lepromatous, L3 If the cases are mixed, as occurs in most instances, they are denoted by both the letters L and N followed by a number to indicate the extent of the lesions

Roentgen Manifestations Since leprosy is rare in the Northern United States, we are indebted to Esguerra Gomez and Acosta and other authors for our knowledge of the roentgen manifestations The early changes are decalcification and rarefaction Simultaneously with the leprous mutilations caused by trophic lesions there is markedly decreased density of the bones in many of the patients In neural leprosy as well as in the lepromatous and mixed types there is frequently present in the epiphyses of the phalanges metacarpals and metatarsals a trabecular structure which is much more differentiated than in healthy individuals The cortical layer of the diaphysis is thin and there is widening of the medullary canal The loss of calcium salts which is manifested by the prominence of the trabeculæ precedes the rupture of some of the trabeculæ and results in the formation of clear circumscribed zones which are located, as a rule, at both ends of the phalanges and the distal ends of the metacarpals and the metatarsals The vacuoles are not accompanied by any peripheral reaction In some instances they distend and destroy the cortex The nutrient foramens are enlarged and become visible on the roentgenograms The decalcifications and rarefactions with multiple vacuoles *occur at any age and in all types of the disease They have been noted in* children in whom ossification is far from complete The changes are usually more apparent in the bones of the hands than the feet The vacuolar rarefactions are due in some instances to nerve damage and in others to the growth of Hansen s bacilli in the marrow and the bone tissue The decalcification in the first phase of osseous rarefaction is due to three factors (1) impaired circulation as the result of deterioration of the sympathetic fibers of the nutrient arteries (2) disorders of the nerve supply because of damage to the conductive nerves and (3) disturbed metabolism of calcium salts In a large percentage of cases there is definite hypercalcemia The rarefaction and decalcification comprise the earliest roentgen manifestations of leprosy

In association with the areas of rarefaction there appear areas of hypercalcification These are frequently associated with a definite increase in the width of the proximal ends of one or more of the phalanges which cover the adjoining phalanx like a cap This appearance has been termed ' hooded It is considered typical of the disease Hyperostosis of the bone occurs in many cases There is increased density in the phalanges metacarpals and metatarsals particularly in the diaphysis contrasting with the rarefaction of some phalanges and normal calcification of others There may be hypertrophy without hyperostosis at the articular ends of the phalanges metacarpals and metatarsals playing a very important part in certain joint deformities which may develop

Leprous reabsorption takes two forms simple reabsorption and reabsorption following atrophy Both are present in cases of pure neural leprosy of grades N2 and N3 and in the mixed forms in which the neural component is of either of these two grades Both types affect the phalanges the metacarpals and metatarsals The changes are more frequent

in the distal phalanges where the process usually begins The reabsorption does not always start in the inner fingers of the hand nor in the outer toes Simple reabsorption occurs at the distal ends of the third phalanges of the hands and feet and there is no associated adjacent hypertrophic reaction or sequestrum formation In some instances there are small erosions at the terminal segment of the phalanx while in others the phalanx appears to be cut across transversely or obliquely The absorption of the phalanges is preceded by onychia The process progresses and invades the proximal end of the phalanx In the early stages there is an increase of the density with expansion of the phalanx producing a type of "hood" similar to that described previously This is presumably a defensive reaction but despite the apparent attempt to arrest the process, the bone is eventually destroyed and the joint becomes involved

Reabsorption following atrophy is a different process and is associated with leprous neuritis It appears in areas supplied by the ulnar nerve in the hand and the external plantar nerve in the foot The more advanced the neural damage the more marked the reabsorption becomes When present in the distal phalanges, the diaphysis becomes thin and cone shaped in its distal half The conical thinning slowly shortens the phalanx with gradual disappearance of the distal portion until only the proximal third remains The para articular portion remains of normal thickness, the rest being very thin The appearance has been described as that of a collarbutton The process continues to advance until only the para-articular portion remains unaffected resulting in a hood type of image Processes beginning in the phalanges of the first and second row produce a different appearance The phalanx in which the cortical layer was previously thinned becomes narrow at its mid-portion and eventually results in a fracture in the mid portion of the phalanx The distal segment disappears and the proximal portion assumes first a collarbutton and later a para articular hood appearance Lesions which involve the phalanges of the first two rows result in the third row losing their normal articulations and becoming deflected with marked distortion muscular atrophy, tendinous retraction and relaxation of the fibroarticular elements In some instances with reabsorption or atrophy of the metacarpals metatarsals, and the first row of phalanges a synostosis may develop between the remaining phalangeal fragments and the atrophic ends of the metacarpals or the metatarsals

The trophic lesions in association with the loss of sensibility cutaneous and vascular alterations and the progressive amyotrophy transform the hands and feet of the leper into stumps These are pitiful in appearance The mutilations may affect all four extremities and are generally symmetrical although frequently predominant in the lower limbs In the hand the reabsorption stops at the carpus while in the foot is is arrested at the tarsometatarsal articulation or may extend to the cuboid In the hand the destructive process is less marked along the outer portion while in the case of the foot the changes decrease in severity from the outer to the inner edge and the appearance may be that of a triangle The perforating plantar ulcer is a cutaneous trophic lesion located on the sole of the foot particularly on the anterior inner arch Because of secondary infection there develops in the late stages a metatarsophalangeal osteo arthritis The roentgen picture is that of osteomyelitis However, the epiphyseal involvement at the first metatarsal and adjoining phalanx

and the destruction of joint structures indicate the diagnosis of leprosy. Healing takes place usually with ankylosis and is associated with varying degrees of deviations of the phalanges and the metatarsals. The leprous whitlow is a true osteomyelitis and occurs with relative frequency in both neural leprosy and the mixed cases in which the neural predominates. On roentgen examination the changes are similar to those of osteomyelitis. The differentiation can be made because there is absence of pain during the early stages of the disease.

Fractures are common not only as a result of reabsorption and atrophy but also because of the trauma to the normal or decalcified bone. The fractures unite with formation of callus. The perforating plantar ulcers cause infectious arthritis as does also the osteomyelitis. In other cases there is an osteoarthrosis which may be of the hypertrophic type with growth at the joints or of the atrophic type associated with thinning of the articular surfaces through leprous absorption. In both instances the position of the bones at the affected joint is altered and deformities occur. The claw hand is caused by amyotrophy of the interossei with predominance of the flexor over the extensor muscles of the hand, hypertrophy of the osseous extremities, and great deviations of the phalanges.

Erickson reports an unusual destructive process of the talus occurring in leprosy. The lesion in the early stages was difficult to detect. There was a small area of decreased density of moth eaten appearance in the inferior portion of the neck and head of the talus. The defect gradually increased in size over a period of several weeks until the entire neck and head of the bone were involved. Cooney and Crosby have described *destruction of the metatarsal and tarsal bones while the phalangeal bones* of the feet remained intact. The changes appear to be absorptive in character. While osteomyelitis, osteitis and arthritis may occur these changes are co existent and are not characteristic of leprosy.

Histopathologic Changes. On histologic examination of the sections of normal phalanges in leprosy there is fraying of the distal ends of the terminal phalanges, concentric atrophy of the phalanges, various other deformities and diffuse osteoporosis. The microscopic study of normal phalanges in these patients may occasionally reveal small gaps in the cortical bone filled with connective tissue. In early neural leprosy the first change is enlargement and increase in the number of the gaps, particularly along the distal margins of the phalanges. The gaps become filled with connective tissue continuous with the marrow and the periosteum. In the later stages with gross deformities and no roentgen evidence of osteoporosis the cortical bone remains dense and of normal width to the distal margin of the bone. In those instances in which there is evidence of osteoporosis the cortex is narrowed and there is usually evidence of osteoclastic activity. In the presence of concentric atrophy, the cancellous bone undergoes absorption and the marrow cavity is narrowed but the cortical bone remains intact. Osteoclastic absorption of bone cannot always be detected probably due to the chronic nature of the process.

ADDITIONAL READING

ESGUERRA GOMEZ G and ACOSTA E Bone and Joint Lesions in Leprosy Radiol *50* 619 1948

MURDOCK J R and HUTTER H J Leprosy a Roentgenological Survey Am J Roent *28* 598 1932

BRUCELLA INFECTION (BRUCELLOSIS, UNDULANT FEVER) OF BONES AND JOINTS

Brucellosis is a generalized infection caused by one of the sub species of Brucella melitensis. The natural habitat of the organism is in domestic animals. In the cow, the disease is characterized by repeated abortions. This is followed in many instances by a carrier state due to chronic infection of the udder and excretion of the bacteria in the milk. The principal sources of contact through which humans acquire the disease are cattle, goats, sheep and swine. The relationship of undulant fever in man and epidemics of abortions in animals was not appreciated until Evans in 1918 stressed the fact that the same organism caused both conditions. The clinical manifestations are indefinite and are frequently so bizarre that the patient is termed a neurasthenic. An early complaint is low back pain. Although the disease is usually termed "undulant fever" there is little or no elevation of temperature in many cases. It may closely resemble tuberculosis, syphilis, typhoid, paratyphoid, rheumatic fever, influenza, and many other diseases. Brucellosis affects the vertebrae more frequently than other bones in the body. The most common site is the lumbar spine. Abscesses usually do not form. Multiple joints may be involved. The arthritic changes develop approximately two or three months after the onset of the disease, an important aid in differentiation of the condition from osteomyelitis or pyogenic arthritis. In a case recently reported by Ploussard, there was involvement of the hip in a male thirty seven years of age. The joint presented the typical alterations of septic arthritis. Re examination four months later showed involvement of the femur with an area of increased radiance in the shaft and localized destruction in the region of the greater trochanter.

Roentgen changes occur in the vertebral bodies and the intervertebral discs, the sacro iliac regions, and other joints. In the early stages, the first manifestation is lack of definition of the bone structures. Osteoporosis and foci of decalcification associated with attempts at bone regeneration are indicative of the chronic stage. The areas of rarefaction in the bone may be small and localized or extensive. Narrowing of the affected joint spaces and destruction of adjacent bones are characteristic manifestations. Signs of bone regeneration may occur in the acute stages and may be associated with extensive spur formations which resemble exostoses. In the spine, extensive calcification occurs within the central portion of the disc and along the anterior ligaments and indicates healing. Destruction of the body of the vertebra and disc may be marked, particularly in adolescents. Signs of new bone formation and repair may occur in areas where no destructive changes had previously been demonstrated. After the changes have been present for several years, the roentgen appearance is that of a chronic degenerative type of arthritis. The joints do not ankylose. The manifestations are very similar to those in tuberculosis and typhoid. Early and extensive bone proliferation aids in establishing the diagnosis. Brucellosis is a chronic disease. The roentgen changes are not pathognomonic and the clinical and laboratory findings must be carefully correlated. The important manifestations comprise joint destructions, multiple areas of involvement, various stages of the disease appearing simultaneously, and a marked tendency to bone repair.

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LYMPHOGRANULOMATOSIS GRANULOMA INGUINALE

The bone manifestations of lymphogranulomatosis are often overlooked during life They have been recorded as occurring in 15 per cent of the cases at autopsy The spine is most commonly affected, next in order of involvement being the sternum, pelvis, femur, ribs, humerus scapula, and clavicle Definite diagnosis may be difficult or impossible and cannot be made without biopsy of the glands Metastatic lesions from other tumors such as Ewing's sarcoma, reticulum cell sarcoma, and similar conditions can produce closely similar osseous changes The roentgen manifestations may consist of a focal process with sharply demarcated borders or a diffusely infiltrating type of lesion The latter is more commonly seen in the acute type of disease while the focal, sharply delineated type is noted in patients with a tendency to a more chronic course The completely osteosclerotic form occurs only in the very chronic types and is frequently associated with involvement of the periosteum Five different varieties of the disease have been described (1) a diffuse osteolytic form (2) a cyst like rarefaction with marginal sclerosis and bridges of normal bone between the individual lesions, (3) a periosteal type with elevation of the periosteum, (4) a diffuse osteosclerotic form which imparts a ground glass density of the bone and (5) the trabeculated, sclerosing form without distinct demarcation The various forms may occur singly, in combination, or successively

Granuloma inguinale is generally a local disease which affects the mucous membranes and skin of the genital and inguinal regions On *occasion it may become more widespread and involve other portions of* the body The fact that the condition may extend to the bone was first noted in 1903 by Hoffman No reports appeared in the roentgen literature prior to 1947 Paggi and Hull described a case in a Negro female twenty-one years old with osteolytic lesions in the medial end of the left clavicle the anterior portion of the right rib and the medial aspect of the right scapula The lesions developed subsequent to a primary lesion in the cervix which had extended to the vulva Donovan bodies were demonstrated in the ulcerated cervix, the labium major, and the material aspirated from the abscesses of the affected bones Lyford, Scott, and Johnson reported three cases with osteolytic lesions in the bones In the first roentgenograms revealed diffuse destruction of all the small bones of the hands and feet areas of osteolysis in the left fibula both tibias, the skull the ulnas the distal end of the right clavicle and the acromion process of the right scapula The lesions were entirely osteolytic without residual sequestrum or evidence of surrounding bone reaction There were destructive changes in the bodies of the fourth and fifth lumbar vertebræ and left hip in the second patient The third case showed osteolytic changes in the left first carpal bone and subsequently destructive changes in all the carpal bones and the distal end of the left radius The changes became so extensive that amputation of the left mid forearm and later at the surgical neck of the humerus became necessary The lesions in the bones contained Donovan bodies

Other observers have reported similar examples In one instance there was a large bone defect in the proximal portion of the tibia at the level of the tubercle The defect measured about 4 cm in diameter and extended from the anterior surface to the posterolateral aspect with extensive dissolution of the cortex The cavity in the bone was surrounded by moderate sclerosis with irregular periosteal thickening in the adjacent portion

of the tibia There was moderate osteoporosis of the distal femur and the tibial condyles A cyst like zone of rarefaction measuring about 1 cm × 1 5 cm was present in the anterior portion of the seventh left rib The rarefied area was demarcated by a zone of sclerosis A case reported by Rhinehart and Bower showed bone lesions in the tibias, the fibulas the ulnas, the upper end of the left radius, the shaft of the proximal phalanx of the right ring finger the frontal bone, and the left astragalus The entire shaft of each tibia was involved, principally along the anterior surface The fibulas showed changes in the distal thirds of the shafts In the astragalus the involvement was along the superior aspect anterior to the articulation with the tibia An area of radiance was present in the lateral aspect of the head of the left radius and the ulnas were involved in their lower halves The skull showed multiple lesions in the region of the suture line between the two frontal bones and in the region of the left frontoparietal suture The diploic cancellous bone between the inner and outer tables was demineralized and there was generalized osteoporosis of the bones of the extremities

The lesions in the bones are multiple, irregular, and destructive with no evidence of sequestrum formation The margins of the defect are indistinct and fade into the normal adjacent bone There is not the punched out appearance which occurs in multiple myeloma and there are no evidences of periosteal proliferation or periostitis The diagnosis is established by aspiration of the lesion and biopsy to demonstrate the characteristic cells of granuloma inguinale with Donovan bodies There is nothing specific in the roentgen manifestations to permit of a definite diagnosis The changes are similar to those which occur in osteolytic or osteoclastic diseases such as metastatic carcinoma leukemia Hodgkin's disease, lymphoma multiple myeloma, and other similar conditions The diagnosis cannot be established without the clinical history physical examination and laboratory studies The location of the primary lesion persistent secondary anemia and severe constitutional reaction with prolonged markedly elevated temperature are important clinical features Disseminated granuloma inguinale is characterized by osteolytic lesions which involve both the long and short bones with no new bone formation as a rule Bone lesions of this type in the presence of obscure genital lesions should suggest the diagnosis

ECHINOCOCCUS DISEASE

Parasitic infestations of the bones are very rare The diagnosis is seldom established by clinical or roentgen methods, particularly in regions where the disease is not endemic The disease is characterized by painless swelling There is usually no evidence of abscess formation and sinus tracts do not develop The ribs vertebræ and other bones may be affected It is a chronic condition which persists for long periods of time Bone involvement occurs in only about 2 per cent of the cases, the spine being the most frequent site of the disease, probably because of the richness of the blood supply

Roentgen Manifestations In the case of the ribs and other flat bones there is usually extensive destruction of bone, only a faint outline of the bony structures remaining Ballooning and multiple cystic changes are frequently present The lesions are multiple and tend to involve adjacent bones by direct extension Echinococcus disease of the vertebræ is manifested by extensive osteolytic lesions with no evidence of compres

sion, wedging, or gibbus The disc spaces are preserved In the later stages there may be evidence of sclerosis and calcification with the formation of a mass The mass may extend paravertebrally in the region adjacent to the bodies of the affected vertebrae The calcification is comprised of lacunae with extensive sclerosis There are areas of bone destruction and sclerosis which are out of proportion to the duration of the symptoms There are no cyst-like areas This is because the disease remains confined to the bone for a long time and progresses very slowly as long as it is so confined Once the bone is eroded there is extraosseous spread in the muscles or the spinal canal which proceeds rapidly and causes symptoms There is no expansion of the vertebra or periosteal reaction The disc may be involved but only late in the disease



Fig 186 Echinococcus Disease of Ilium There are extensive cystic changes with dissolution and multiple pathologic fractures of the ilium

Differential Diagnosis In the differential diagnosis it is necessary to include tuberculosis other parasitic diseases bone cyst giant-cell tumor, osteitis fibrosa cystica Hodgkin s disease multiple myeloma and metastatic carcinoma Hemangioma may produce a closely similar picture In tuberculosis the involvement tends to be less extensive and the intervertebral discs are affected Giant cell tumor and bone cyst are slow growing expand the bone and do not involve contiguous structures In osteitis fibrosa cystica Hodgkin s disease multiple myeloma and metastatic carcinoma the clinical manifestations aid in establishing the diagnosis As with other rare and unusual conditions it is essential to bear the possibility of the disease in mind if the correct diagnosis is to be made The presence of eosinophilia positive complement fixation test and the history indicate the correct diagnosis The demonstration of the parasites by puncture biopsy establishes the diagnosis with certainty

Treatment Radiation therapy results in bone repair calcification and sclerosis of the cyst There is usually return to nearly normal function of the spine

Chapter

5

Glandular, Nutritional, and Metabolic Disturbances

THE ENDOCRINE GLANDS

THE normal development, growth and function of the human body are orderly in time and space and are genetically determined, due in large degree to the safeguard afforded by a system of ductless glands termed the endocrines or the glands of internal secretion The endocrine glands elaborate specific products which are secreted into the blood stream and are distributed from the place of origin to every part of the body The products are termed hormones because they set in motion cellular activities in distant organs It must be stressed that the hormones which act as catalysts are not uniformly stimulating in their effects in some instances acting in such manner as to inhibit the activities of cells Since the hormones are transported throughout the body the presence of one hormone can influence the activity of one or more of the other endocrine glands Proper interaction of the ductless glands is known as endocrine balance The interaction of the various glands has made the study of endocrine function extremely difficult since interference with or removal of one of the glands influences other components of the endocrine system The blood stream furnishes the means of transportation for the hormones hence hormone therapy is most effective after injection of the hormone Relatively few of the hormones are effective on oral administration Extracts of the gonads, the thyroid, and the adrenal cortex may be given orally

The hormones are not specific in action in different mammals and it is possible to utilize hormone therapy on a large scale by the administration of extracts of non human organs, particularly cattle and sheep The sex hormones and a few others have been synthesized The use of these synthetic products is preferable to the administration of extracts of an entire organ which contains factors other than the active hormone Disturbances of endocrine balance are in the main due to quantitative changes in certain hormones Whether qualitative changes can occur in the hormones is not known Excessive production of hormone is due to hypertrophy or hyperplasia of a ductless gland or a tumor with proliferation of the functional elements of the gland Deficiency in the production of a hormone is caused by atrophy or degeneration of the glandular cells Destruction of the glandular elements may be caused by a tumor with proliferation of the interstitial elements of the gland or a metastatic tumor within the gland In rare instances there is congenital absence of the endocrine gland An important example is absence of the thyroid

gland in cretins The endocrine glands which are known to influence the bones are (1) the pituitary gland or the hypophysis cerebri (2) the thyroid gland, (3) the sex glands or gonads, and (4) the parathyroid glands

THE PITUITARY GLAND OR HYPOPHYSIS CEREBRI

The hypophysis or the appendage of the brain is situated in the hypophyseal fossa of the sphenoid bone It is comprised of two principal parts an oral and a neural portion The neural segment develops from the floor of the diencephalon behind the optic chiasm, its connection to the brain persisting as the hypophyseal stalk The oral portion develops as an epithelial pouching on the roof of the primary ectodermal oral cavity or Rathke's pouch and embraces the neural lobe anteriorly and laterally This part loses its connection with the pharyngeal epithelium at an early stage of development The oral segment is subdivided into three portions (1) the anterior lobe proper or the glandular lobe also termed the distal lobe (2) the pars intermedia, and (3) the pars tuberalis The anterior lobe of the pituitary gland is the site of production of the hormones which either directly or indirectly exert an influence on the skeleton Many hormones are elaborated by the pituitary gland However, only four are of immediate interest (1) the growth promoting or growth hormone (2) the gonadotropic hormone (3) the thyrotropic hormone, and (4) the adrenocorticotropic hormone The growth hormone acts as a direct stimulant while the thyrotropic hormone affects skeletal growth indirectly The effects of the gonadotropic and adrenocorticotropic hormone are antagonistic the result being inhibition of growth The production of only two of these hormones is ascribed to the activity of certain cells of the pituitary gland the eosinophilic cells producing the growth hormone and the basophilic cells producing the gonadotropic hormone It is not possible to observe the function of the endocrine glands directly The activity of the glands is dependent on experimental and clinical observations and must under normal conditions be determined by indirect methods Hormonal deficiency or hormonal excess is produced experimentally or is observed in the patient and in this manner the activities of the glands are deduced An understanding of the influence of the hypophysis on growth has been attained by a study of patients in whom a tumor or hypertrophy of the eosinophilic cells has led to an overproduction of the pituitary growth hormone In these instances, the growth potential of the body and various organs is increased In consequence there is a more or less proportional or in some instances extremely disproportional overgrowth Whether one or the other will result is dependent on the time of onset of the disease

The pituitary cells are classified according to their staining qualities The functional cells or glandular part are of three types (1) the chief cells or chromophobe cells these comprise 52 per cent of the cells and stain poorly (2) the basophilic cells comprise 10 per cent and are also termed cyanophilic or beta cells and (3) the eosinophilic cells known also as acidophilic oxyphilic or acid cells

(1) **Chromophobe Adenoma** The chromophobe adenoma produces a bulky tumor of the pituitary with resultant enlargement of the fossa and hypofunction of the gland due to pressure There are no characteristic roentgen manifestations except those in the sella

(2) **Basophilic Adenoma** Basophilic adenoma is associated with Cushing's syndrome, a rapidly acquired, painful adiposity of the face, neck and trunk. There are no changes in the extremities. Amenorrhea and impotence develop. In the female there is hypertrichosis of the face and trunk, hypertension, backache, abdominal pains, easy fatiguability, and marked weakness. The bones show very marked generalized decalcification, especially in the pelvis, spine and skull. The vertebrae show wedging. Spontaneous fractures may occur.

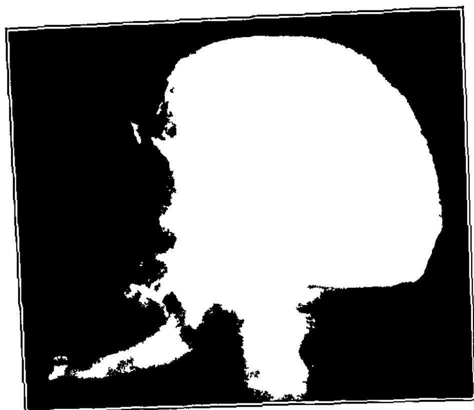
(3) **Eosinophilic Cell Adenoma** (*a*) *Acromegaly* Hyperpituitarism developing during adult life results in acromegaly. The disease is characterized by hyperfunction of the anterior lobe of the pituitary gland with usually an adenoma of the eosinophilic type. It occurs in both sexes with about equal frequency. In contrast to gigantism, the hyperpituitarism of adolescence and youth, the condition is characterized by the development of striking disharmonies and disproportions of the body. Hypersecretion of the pituitary growth hormone in the adult acts as a powerful stimulant. Many internal organs enlarge and the skeleton also attempts to respond to the stimulus. However, the skeletal response is limited by the fact that the principal mechanism of longitudinal growth, the epiphyseal plate, has already disappeared.

The manifestations of acromegaly are well known and present a classical picture. There is enlargement of the nose, fingers and toes, the so called acral parts of the body. The enlargement of the nose is due principally to the growth of its cartilaginous skeleton with overdevelopment of the septal cartilage, the triangular cartilage and the alar cartilage. There is an associated hypertrophy of the overlying subcutaneous and cutaneous tissues, which contributes to the enlargement of the nose and the lips. Overdevelopment of the masticatory skeleton ensues with enormous enlargement, disproportion and protrusion of the mandible. The teeth are widely spaced. There is an extreme degree of thickening of the bones of the skull with overgrowth of the ridges. The sella usually shows characteristic changes and becomes enlarged in a globular fashion. The clinoids may be absorbed and the floor thinned, depressed or destroyed. The nasal accessory sinuses are greatly enlarged. The ribs become heavier than normal and are elongated in both the bony and cartilaginous portions. The curvature of the ribs is decreased and the chest becomes bovine in type with a great increase in the anteroposterior diameter. The changes in the thorax are accounted for by elongation of the ribs at the junction between the bone and the cartilage since endochondral ossification continues during adult life in this region, the only area in which the ribs of an adult can continue to grow. There is massiveness of all the bones, the long bones becoming markedly thickened and the bodies of the vertebrae enormously enlarged. Proliferation of the articular cartilages results in appositional growth of the epiphyseal parts of the tubular bones by endochondral ossification and arthritic changes. Elongation of the hands and feet and hyperostosis of the terminal phalanges ensue. The fingers and toes become elongated due to the fact that each is composed of several bones and the deposition of small amounts of bone at all of the articular surfaces produces a true increase in the length of the digits. The growth of the bodies of the vertebrae in acromegaly occurs almost exclusively by periosteal apposition of lamellated bone at the lateral and anterior surfaces of the body. The intervertebral disc increases in the transverse and anteroposterior diameters to the same extent as the

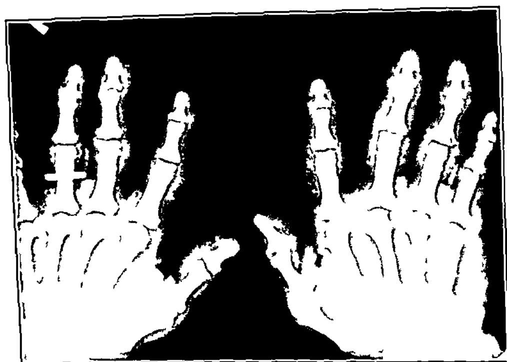
vertebra The fibrocartilage differentiates into hyaline cartilage particularly at the periphery of the disc in the region adjacent to the bone In consequence a new site of endochondral ossification is formed with the resultant development of spurs and shelf like hyperostoses at the margins of the vertebral bodies

The changes in the skull associated with acromegaly vary widely However, they usually fall into a definite pattern Deviations from the pattern are dependent on factors such as the sutural age of the individual at the time of the onset of the disease and similar variables The features which play a determining role are those of the facial skeleton The most pronounced changes are an enormous growth of the cranial superstructures with marked prominence of the supraorbital, occipital, and zygomatic arches and the mastoid region The external occipital protuberance becomes very prominent and in some instances the occipital bone at this point measures 4 cm in thickness The diploe consists of dense, regular arranged spongiosa The mastoid process is increased in all dimensions so that the greatest width of the skull occurs between the most prominent part of the mastoid processes There is marked widening of the orbits laterally The increase in facial height is contributed to almost equally by the maxilla and the mandible The development of the skull in the first years of life normally proceeds from a preponderance of the neurocranium to a relatively greater increase in the size of the masticatory part and during this period the cranial superstructures develop In some instances the abnormalities of the acromegalic skull appear as an exaggeration of the early growth changes and result in a grotesque appearance It must be stressed that in the development and growth of the cranial superstructures there is practically no sutural growth, the process proceeding by simple surface apposition of bone In consequence hyperfunction of the hypophysis results in changes during adult life which are similar to those which take place during the period of normal growth The mandible normally grows by endochondral ossification at the condyles and surface apposition in certain areas In acromegaly the growth of the mandible is again initiated and continues at a time when it normally has ceased The changes in the maxilla differ from those in the mandible because the growth of these bones occurs in different manners In the maxilla sutural growth plays the principal role as regards the transverse and anteroposterior planes As a rule sutural growth has ceased when the acromegalic changes are initiated In consequence the maxilla remains narrow and short and may maintain its normal relation to the cranial base The height of the alveolar process becomes increased The growth of the mandible and maxilla may be asymmetrical due to the fact that one condyle of the mandible is shifted laterally In the maxilla one alveolar process may accentuate the asymmetry by growing vertically rather than downward and slightly upward

(*b*) *Gigantism* When overproduction of the growth hormone occurs in childhood or during the period of adolescence that is at a time when generalized growth of the individual is still progressing there results an exaggeration in size of the genetically fixed pattern This is primarily due to the fact that all of the structures necessary for normal growth are still present and active in the patient Two of these structures merit special attention the epiphyseal plates and the sutures of the skull The sutural growth of the neurocranium or the brain case is dependent on the growth of the brain It is known that the brain does not undergo exces



A



B

FIG 187 Acromegaly The Skull A A characteristic acromegalic appearance with well developed sinuses and enlargement of the bones of the skull particularly the base the face and jaws The angle of the mandible is increased the jaw juts forward and there is lack of occlusion of the teeth The bony ridges of the skull are thickened and prominent The pituitary fossa is large the floor is irregular and intact There are no destructive changes in the dorsum or the clinoids B The Hands The bones of the hand are short and dense There is marked widening of the tufts of the terminal phalanges of the fingers The soft tissues of the distal portions of the fingers are thickened and increased in size

sive growth in cases of pituitary tumor which develop during youth. As there is no increase of the stimulus normally exerted by growth of the brain, the neurocranium remains normal in size. This is of extreme importance as it proves that the presence of the growth hormone alone does not suffice to promote growth in all parts of the body. That there is no enlargement of the brain is explained in part by the fact that the hypophysis has little or no influence on ectodermal structures. The dependence of neurocranial growth on that of the brain causes loss of the proliferative power of the sutural connective tissue after the brain has attained its normal volume. This faculty once lost cannot be restored by activity of the hypophysis. The epiphyseal plates are the main sites of the longitudinal growth of the trunk and the extremities are stimulated to abnormal activity. This activity persists for a long period of time. In consequence, the development during early life of an eosinophilic adenoma of the hypophysis results in an increase in the height of the body far beyond normal and giants with a height of 7, 8, and even 9 feet have been known to develop. Some authorities are of the opinion that epiphyseal growth, which comprises primary growth of cartilage, is affected principally. Simultaneously with the exaggerated longitudinal growth in gigantism the skeleton also shows increased transverse growth of the tubular bones and marked thickening of the bones of the skull.

The proportions of the human body are not markedly distorted in cases of gigantism. However, an absolute maintenance of the normal proportions is not always found. This is due to the fact that some parts of the body complete their growth so soon after birth that even the onset of the disease relatively early in life can no longer exert an influence upon them. The brain and higher sensory organs are in this category and a disproportion between the skull and the face necessarily results. In consequence of this lack of growth of the cranial skeleton, there is a disharmony between the upper and lower jaws. Lack of growth of the skull may be partially compensated for by thickening of the bones of the skull. In the case of the jaws, there is a marked disproportion between the size of the crowns of the teeth and the jaw bones. This is due to the fact that the teeth cannot enlarge, their size being determined in early life, and also because the growth hormone apparently does not influence epithelial growth. Since some of the epiphyseal plates of the various bones ossify soon after puberty, while others persist to the twentieth or twenty second year of life, there are numerous other instances of minor variations and disproportions of limbs and parts of limbs if the tumor develops during the years prior to epiphyseal closure.

Hypopituitarism. Pituitary Dwarfism. Pituitary dwarfism is termed nanosomia pituitaria. The proportions of the skeleton of a pituitary dwarf are normal in so far as they represent the proportions of the infantile or childhood type of skeleton which prevailed at the time of onset of the disease and account for the skull and head being proportionately large. The eruption of the teeth is retarded. An important feature of hypopituitary dwarfism is persistence of the epiphyseal plates and synchondroses to old age. This contradicts the fact that growth has ceased. However, the cessation of growth is of a different order from the cessation of normal growth. Normally the cartilage ceases to proliferate while its resorption by connective tissue elements and its replacement by bone proceed until synostosis of the bony elements, the epiphysis and the diaphysis, has been accomplished. In cases of pituitary deficiency, growth

of cartilage and bone are arrested simultaneously The epiphyseal plates persist because bone growth has been interrupted In pituitary dwarfism, the major change in the region of the epiphyseal plate is the development of a thin, compact lamella of bone termed the terminal plate which results in sealing the marrow spaces of the metaphysis against the cartilage

Deficiency of the growth hormone is usually the result of destruction of the anterior lobe of the hypophysis and is due to the presence of an extrahypophyseal tumor such as a craniopharyngioma, a cyst of the hypophysis, or tuberculosis of the pituitary gland In rare instances there may be idiopathic deficiency of the eosinophilic cells While hyperpituitarism leads to overgrowth hypopituitarism is characterized by the reverse and results in the development of pituitary dwarfism The clinical manifestations are dependent on the time of onset of the destructive or degenerative changes in the pituitary When the onset of hypopituitarism takes place at an early age, there is generalized cessation of growth with proportionate dwarfing of the entire body If the condition is associated with degeneration of the eosinophilic cells of the hypophysis at the end of the growth period, disharmonies result because the distal parts of the extremities are affected more than the proximal portions and acromicria ensues *Acromicria* is the counterpart as regards symptomatology and etiology of acromegaly It is a rare condition and is characterized by generalized slenderness of the bones with rarefaction of compact and spongy bone, smallness of size of the nose and facial skeleton and shortness of the extremities The roentgen changes are not characteristic and cannot be differentiated from other forms of retardation of growth The bones resemble those of a young child

THE THYROID GLAND

The thyroid gland develops from an epithelial budding on the floor of the primary oral cavity It consists of spheric or ovoid follicles lined with simple cuboidal epithelium and filled with an eosinophilic colloid There is an important interdependence between the thyroid gland and the hypophysis The hypophysis produces the thyrotropic hormone, the presence of which is indispensable for the activity of the thyroid gland Lack of this hormone accounts for the involution of the thyroid gland after hypophysectomy The thyroid hormone, the iodine containing thyroxin, is indispensable for the maintenance of the normal metabolic rate of the tissues A knowledge of the interrelationship between the thyroid and pituitary glands is essential in order to understand the disturbances of growth which result after thyroidectomy and aplasia or hypoplasia of the thyroid gland Because of the thyroid deprivation there develops thyroprival or secondary pituitary dwarfism and this is complicated by the direct effects of thyroid deficiency metabolism on the cells Pituitary or thyroid dwarfism of various types ensue dependent upon the time of onset of the primary disturbance The most severe changes are observed in cretinism in which the thyroid gland is congenitally absent Cretins may attain a body height of 70 to 100 cm A second form of this condition is termed infantile myxedema and is due to degeneration of the thyroid gland in early childhood Individuals with this lesion may attain a height of 100 to 150 cm Pituitary dwarfism with

the effects of destruction of the hypophysis become manifested at puberty and the height of the patient varies from 130 to 160 cm The final stage is acromicria in which the anomaly develops during the gradual termination of skeletal growth Neither hypopituitarism nor hypothyroidism developing in the adult causes gross changes The analogy between pituitary and thyroid dwarfism holds true only for the skeleton and the development of the teeth In other respects hypopituitarism and hypothyroidism differ markedly in their manifestations The most striking difference is concerned with the mental ability, which remains unimpaired in the pituitary dwarf but is markedly retarded in the thyroid dwarf The patients in the two conditions present strikingly different appearances The pituitary dwarf usually is normally proportioned The thyroid dwarf is myxedematous and tends to be obese with firm edema of the subcutaneous tissue Hypogonadism occurs both in pituitary and thyroid deficiency

Cretinism Cretinism is apparently the result of interference with normal embryonic development of the thyroid It is more common in females The condition frequently is unrecognized until the child is several months old There is coarsening of the features thickening and enlargement of the tongue and retardation of physical and motor development The hair is dry The stunting of the patient s growth is due to interference with normal development of the epiphyseal cartilages with resultant irregularities of ossification *The characteristic roentgen manifestation is stippling of the epiphyses* In the cretin the proportions of the skeleton and the body are those of an infant The head is excessively large in proportion to the body and the cranial skeleton is relatively larger than the facial skeleton The synchondrosis at the base of the cranium and the sutures of the skull remain open The teeth are retarded in development and in eruption although the size is not altered In consequence the teeth and the alveolar process appear large in proportion to the maxilla and mandible The bodies of the vertebræ and consequently the vertebral column are short The intervertebral discs may become as thick as the vertebral bodies The ribs are slender In the sternum there is characteristically a persistence of cartilage The limbs are short with relation to the length of the trunk This characteristic is also present in the body of the infant The epiphyseal cartilages and the cartilage of the synchondroses in the cranium and face persist After the proliferation of cartilage has terminated practically completely the proliferation of the connective tissue in the adjacent marrow spaces also ceases There is apposition of bone probably the result of adaptation to functional stimuli This produces a terminal plate in the region adjacent to the cartilage which seals the marrow spaces of the epiphysis and the diaphysis Consequently the epiphyseal cartilage and the adjacent bone in the cretin are apparently identical to those in the pituitary dwarf Certain portions of the bones may remain cartilaginous for a long period of time and in the scapula the acromion the coracoid process the vertebral border and the clavicle the ends may consist of cartilage in individuals who have attained the age of fifty or more years In juvenile myxedema the changes in the skeleton comprise an arrest of growth at an age somewhat greater than the critical age in the cretin as in these periods the hypothyroidism develops between the sixth and twelfth years of life

In hypothyroidism growth is not immediately arrested rather it is retarded The final phases of cartilaginous growth are characterized by

irregular arrangement of the multiplying chondrocytes These alterations are manifested roentgenographically by fraying of the metaphyseal lines The presence of multiple centers of ossification is a characteristic finding on roentgen examination These multiple centers of ossification fuse slowly with the resultant development of an irregular epiphysis, the process being termed *epiphyseal dysgenesis* Absence or deficiency of the thyroid gland can be corrected by the oral administration of thyroid extract or the injection of thyroxin The success of the treatment depends to a large degree on the initiation of therapy early during the course of the disease With adequate treatment, the children grow at a normal rate and in some instances there is acceleration of growth until practically normal height has been attained The fact that the increase in growth of the skeleton can be brought about by the injection of pituitary growth hormone proves that absence of the thyroid gland retards the growth of the skeleton through the mediation of the hypophysis In myxedema the mental retardation and depression of the metabolic rate can be eliminated practically completely by the administration of thyroid hormone

In juvenile hypothyroidism there occur delayed growth of bone and dwarfed stature and these manifestations are susceptible of correction by the administration of thyroid extract Overdosage or excessive prolongation of the therapy results in overcorrection with acceleration of ossification and resultant early epiphyseal union which brings growth to a standstill In order to utilize the ossification centers as a guide to thyroid dosage, it is essential that comparative roentgen studies be performed every three to six months and a ratio established between the x ray manifestations and the height measurement as skeletal development is usually in advance of growth in these circumstances Roentgen studies are of particular value to prevent overdosage Hypothyroidism is one of the most frequent endocrine disorders of childhood The condition may be congenital or acquired In the great majority of cases in children, the condition appears to have been present since birth However acquired hypothyroidism may occur in children who have been entirely normal during the early years of life

Angular dorsolumbar kyphosis constitutes a previously unrecognized skeletal sigh of myxedema Swoboda reports 7 cases of infantile myxedema in which gibbus occurred The anomaly was found to be due to varying degrees of wedging of the second lumbar vertebra and in some instances one or two of the adjacent vertebræ In the lateral projection the involved vertebræ show a step-like defect in the superior and anterior aspects which imparts a wedge shaped appearance The roentgen pattern is characteristic In the differential diagnosis it is necessary to include gargoylism (dysostosis multiplex or Hurler's syndrome), Morquio's disease (chondro osteodystrophy) and achondroplasia (chondrodystrophy)

Hyperthyroidism Increased function of the thyroid gland is termed toxic goiter Graves' disease or Basedow s disease and in young individuals may lead to an acceleration of skeletal growth The alterations are dependent on the action of the thyroid hormone on the differentiation and functioning of the eosinophilic cells of the pituitary gland This indicates that hyperthyroidism causes a secondary hyperpituitarism and is associated with increased production of the growth hormone When the overproduction of thyroid hormone exceeds certain limits, toxic symptoms ensue Toxic goiter in the adult may cause simple osteoporosis or a mild degree of osteitis fibrosa In hyperthyroidism enlargement of the para

thyroid gland is present in only a very small number of cases and to only a very slight degree. The bone changes in hyperthyroidism are due to the ensuing acidosis or may be caused by the influence of the thyroid hormone on the kidneys with resultant increase in the excretion of phosphates and calcium by the kidneys. The osseous alterations are the result of the depletion of calcium and phosphorus in the bones. Reduced bone apposition and increased osteoclastic bone resorption in toxic goiter constitute an attempt of the body to mobilize calcium and phosphorus in sufficient amounts to compensate for the increased loss of these elements. Hyperthyroidism is uncommon in childhood, especially in patients less than five years of age. The symptoms and clinical manifestations are similar to those in the adult. There is an increase in height and skeletal development after the disease has persisted in a child for a prolonged period of time.

THE MALE AND FEMALE SEX GLANDS

Both the testes and the ovaries function as endocrine glands as well as organs which produce mature sex cells In the male, the endocrine portion of the organ is represented by the interstitial cells of Leydig The endocrine function of the ovary is performed principally by the epithelial cells of the follicles The sex hormones of the male comprise the andro gens, androsterone, and testosterone and are comparable to the estrogens The primary sex hormones of the female include follicular hormones, theelin, and theelol The sex hormones are produced in greater quantity at the time of maturity and this period in many animals coincides sometimes with an accelerated period of growth The male sex hormone and the primary female sex hormone act as inhibitors of the growth promoting function of the hypophysis The interrelationship between the hypophysis and the gonads is complex The gonadotropic hormone of the hypophysis is produced by the basophilic cells and is indispensable for normal development, particularly the maturation of the sex glands The sex hormones in turn inhibit the action of and eventually cause degeneration of the eosinophilic cells of the hypophysis which produce the growth hormone Thus the growth promoting function of the hypophysis is self-limited, the basophilic cells of the pituitary gland limiting the function of the eosinophilic cells by way of the gonads

Hypogonadism The consequences of removal or absence of the sex glands have been known for thousands of years Castration of domestic animals results in changes in behavior and bodily development It was at first believed that castration and spaying produced feminine characteristics in the male and masculine characteristics in the female This is not the case as after these operations the sex approaches the neutral The changes in the skeleton are numerous and varied There is postponement of skeletal maturity and suppression of secondary sex characteristics The epiphyseal cartilage continues to proliferate far beyond the time of normal closure The resultant overgrowth of the skeleton results in a type of gigantism characterized by extremities which are long in comparison to the trunk It must be stressed that the variations in body proportions may occur under various conditions and that maturity is *normally* reached at different ages by different groups Thus, it is known that girls menstruate earlier in an urban than in a rural population and the latter show proportionately longer limbs than the former *In the skull* of castrated individuals, the continuation of sutural growth, as in other forms of gigantism, is restricted to the facial skeleton, the mandible continuing to grow beyond the normal time There is *no enlargement* of the brain case as growth of the skull is entirely dependent on enlargement of the brain The discrepancy between facial and cranial growth results in prominence of the masticatory apparatus and overdevelopment of the superciliary arches The pelvis after castration or spaying retains an infantile form with the characteristics of the *period* prior to the maturation of the sex glands The changes in the human skeleton are similar in individuals who have been castrated or lost the sex glands because of disease in early life and those suffering from *congenital* hypogonadism

Hypogonadism or eunuchism may result in delay in fusion of the epiphyses If there is simultaneously normal or increased activity of the pituitary growth hormone there may result a marked overgrowth of the long bones By the administration of stilbestrol the rate of bone growth may be increased Whether this results in earlier fusion of the epiphyses or not is uncertain

Hypergonadism Hypergonadism in humans is restricted to precocious development of the sex glands In animals the condition can be produced experimentally by the injection of sex hormones Precocious puberty is due to production in the early months or years of life of sex hormone in sufficient quantity to effect maturation It may be due to primary hypertrophy and hypersecretion of the sex glands In most cases the overdevelopment of the testes or ovaries is secondary to a tumor of the adrenal cortex or the pineal body Tumors of the sex glands may also cause hypergonadism The skeletal manifestations comprise accelerated growth and maturation A child of five years may attain a body height of 140 cm Roentgen studies show that the ossification corresponds to an age of ten or more years greater than normal Despite the accelerated and precocious growth, the individual remains small apparently due to premature closure of the epiphyseal plates These findings contrast sharply *with the changes in hypopituitary and hypothyroid dwarfs as in* these conditions the epiphyseal cartilages persist to old age The changes in growth in hypergonadism are best described as a distortion and exaggeration of the normal pubertal spurt of growth which leads to termination of longitudinal growth

The Action of the Secondary Female Sex Hormone or Progestin The hormone of the corpus luteum, which is indispensable for normal gestation is in many ways antagonistic to the effect of estrin, the primary hormone This *antagonism is best illustrated by the changes in the skeleton during* pregnancy In young women, there may be a spurt of growth during pregnancy This is possible only during the time when the epiphyseal cartilages are present After the cessation of general growth skeletal changes related to the action of the secondary female sex hormone are limited to the pelvis These are twofold First, there is enlargement of the pelvic canal in all diameters Secondly, there is relaxation of the pelvic ligaments The growth of the pelvis occurs particularly at the symphysis and is due to simultaneous proliferation of the symphyseal cartilage and apposition of bone at the pubic bones These changes are not dependent on the age of the individual There is also growth at the sacro iliac articulation the cartilages of the articular surfaces of the sacrum and ilium becoming the sites of endochondral proliferation

The Relationship of Estrogens and Bone Formation in the Human Female In postmenopausal osteoporosis there is a hormonal deficiency and it is possible to cause calcium retention in the human body by the injection of estrogens The metabolic effects are accompanied by a concomitant improvement in symptoms Osteoporosis is a disorder in which the etiologic factor is a hypofunction of the osteoblasts in the production of bone matrix Although the mechanism of the reaction is not known, it appears that the estrogens stimulate the osteoblasts so that the process of ossification proceeds normally However it has not been definitely shown that bones become denser under treatment although in some instances it has been reported that there is actual reconstitution of bone Withdrawal of the estrogen produces an exacerbation of the condition while further administration results in improvement again There is no evidence on clinical or roentgen study of any tendency of the bone to undergo absorption except when the estrogens are discontinued In Paget's disease of the bone there is increased circulation Therefore, it appears probable that the ordinary process of postmenopausal atrophy developing in a bone which is the site of an active Paget s disease is greatly augmented both in rapidity and degree

DISTURBANCES OF THE PARATHYROID GLANDS

The parathyroid glands were discovered less than one hundred years ago These glands in the human being comprise four or more small bodies, usually arranged in pairs, at the posteromedial surface of the thyroid gland The size of each gland is about 2 × 4 × 6 mm The lower pair is slightly larger than the upper pair They are yellowish red, the color serving to differentiate them from the thyroid gland One or more of the glands may be embedded in the substance of the thyroid gland and in this manner not be observed during operation or at autopsy The parathyroid glands develop from the third and fourth branchial pouches During their development, they shift in such a way that the third parathyroid becomes the lower gland and the fourth the upper The cells of the parathyroid glands are divided into two types The more numerous are the chief cells Less numerous are the eosinophilic or oxyphilic cells which develop after the tenth year of life After injection of the parathyroid hormone which is known as parathormone, a series of changes ensues First, the re absorption of phosphorus in the kidney is decreased so that the organism is depleted of phosphorus Second, extensive osteoclastic resorption of bone begins Third, the calcium level in the blood rises The correlation of these findings has been a difficult matter The following is the most satisfactory hypothesis The function of the parathyroid is to maintain optimum calcium blood levels This is accomplished by mobilizing calcium from the skeleton In the living organism, extraction of calcium from the body is impossible and immobilization of calcium can be accomplished only by resorption of bone The parathyroid glands are responsible for the differentiation of osteoclasts and the ensuing resorption of bone During the resorption of bone, not only calcium but also phosphorus is mobilized The unphysiologic rise of the phosphorus blood level is counteracted by the function of the parathyroids This is accomplished by a direct influence on the kidney, re absorption of phosphorus in the tubules being inhibited and consequently the excretion of phosphorus becoming increased

The two functions of the parathyroid hormone increased bone resorption and increased excretion of phosphorus, are inseparably linked The fact that after the experimental injection of parathormone the rise in phosphorus excretion precedes by several hours the differentiation of osteoclasts is easily understood The action of the parathyroids on the bone and the kidney is initiated simultaneously The effect on the bone requires time for its development because osteoclasts have to differentiate from osteoblasts in order to become active The effect on the kidney constitutes a functional change in the tubular cells and this takes place almost immediately Observations in cases of hyperparathyroidism have shown that the parathyroid hormone has an additional function which tends to maintain the calcium blood level The parathyroid hormone apparently inhibits the calcification of newly formed bone and prevents precipitation of the calcium liberated through bone resorption This action of the parathyroid hormone is antagonistic to the action of vitamin D which promotes calcification

Primary Hyperparathyroidism Generalized Osteitis Fibrosa

First described by von Recklinghausen in 1891 this condition is also known as osteitis fibrosa cystica and primary parathyroidism It is

associated with increased parathormone secretion due to malignancy, hypertrophy, or adenoma of one or both of the parathyroid glands The disease generally occurs in the middle aged and is about three times more frequent in women than in men The pathognomonic features consist of marked osteoporosis and generalized decalcification of the skeleton associated with alterations in the blood chemistry and calcium balance The bones are porous and filled with osteoclasts and osteoblasts Multiple giant cell bone tumors (osteoclastomas) form with epulis of the mandible and giant cell tumors of the other bones There is very marked, generalized decalcification and skeletal shortening The vertebræ are narrowed and of the fish-type with marked kyphosis Fractures result from very slight traumatisms and from muscle pull without trauma

Askanazy in 1904 was the first to suggest that there might be a relationship between the parathyroid glands and decalcification of the skeleton In 1905, MacCallum called attention to the fact that there is an apparent association between a tumor of the parathyroid gland and chronic renal disease, the renal disease being overshadowed by the skeletal changes Erdheim in 1907 commented on the frequency of hypertrophy of the parathyroid gland in cases of osteomalacia In 1915 Schlagenhaufer indicated that the disease was probably due to enlargement of the parathyroid glands This was in distinct contradiction to the view prevailing at the time that the enlargement of the glands was merely a secondary manifestation However the honor of conclusively establishing the relationship between adenomas of the parathyroid and bone decalcification belongs to Mandel In 1905, he removed surgically an adenoma of the parathyroid and alleviated the metabolic dysfunction in a patient with hyperparathyroidism

Clinical Manifestations The signs and symptoms of hyperparathyroidism are very variable The general manifestations comprise weakness, loss of appetite loss of weight muscle and joint pains, constipation, abdominal pain, bradycardia, cardiac irregularities polydypsia and hypochromic anemia In the skeletal system there occur generalized decalcification cysts and giant cell tumors fractures deformities, shortenings and epulides The manifestations in the urinary tract comprise polyuria albuminuria dysuria hematuria cloudy urine gravel, renal urethral and vesical calculi renal colic diminished renal function and nephrocalcinosis Metastatic calcifications are common and there frequently occur arterial calcification broncholithiasis pulmonary calcinosis generalized calcinosis hypercalcemia hypophosphatemia and hypercalcuria A decrease in serum or plasma phosphates is an important aid in diagnosis The symptoms vary widely in the individual case depending upon whether the skeletal changes of osteitis fibrosa cystica, generalized decalcification of the skeleton without cysts or giant cell tumors or deposition of the calcium in the soft tissues particularly in the kidneys predominate The early symptoms of the disease may permit of the establishment of the diagnosis The first skeletal manifestations comprise pain in the back and extremities muscle weakness pathological fractures localized swelling gross deformities and disturbances of gait There is polyuria polydypsia and colic referred to the urinary tract Marked weight loss occurs Most cases of hyperparathyroidism occur in middle adult life However instances have been reported in which cysts of the jaw and calvaria were present at the age of six months





FIG 188 FIG 189

FIG 188 Osteitis Fibrosa Cystica The middle and lower portions of the humerus show markedly increased radiance multilocular cystic changes expansion and thinning of the cortex The trabecular pattern is obliterated The patient had a parathyroid adenoma

FIG 189 Osteitis Fibrosa with Pathologic Fracture There are multiple cyst formations in the humerus clavicle and ribs with a comminuted fracture of the humerus There is marked generalized osteoporosis

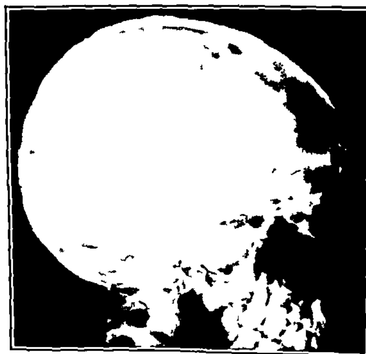


FIG 190 Generalized Osteitis Fibrosa There are multiple discrete areas of increased radiance scattered irregularly throughout the skull There is generalized osteoporosis and mottling of the bones

Hypercalcemia is an important finding in suspected cases of hyperparathyroidism and is an aid in diagnosis, particularly in association with other signs and symptoms of overactivity of the parathyroid bodies This finding is not pathognomonic as the level of serum calcium may be increased in other diseases particularly multiple myeloma and polycythemia vera Also normal values for serum calcium have been found in proved cases of hyperparathyroidism In the healthy person about 70 to 90 per cent of the excreted calcium is in the feces and only small amounts in the urine In hyperparathyroidism the reverse holds true unless large amounts of calcium are ingested

Etiology The etiology of hyperparathyroidism is unknown It has been postulated that certain cells which maintain an embryonic capacity for proliferation are present in certain individuals and create a potentiality for the formation of adenomas Whether a parathyroid adenoma develops in the individual patient retaining cells of this type is dependent upon the intensity of stimulation to which the cells become subjected by lack of vitamin D There appears to be a relatively high incidence of hyperparathyroidism in England and New England as vitamin D deficiency is favored by fog smoke haze in the atmosphere and indoor living habits Many theories have been advanced to explain the mechanism by which the bones become demineralized The best explanation is that of Shelling He is of the opinion that the parathyroid hormone increases the solubility of calcium phosphate in the blood and causes a shift in equilibrium between the solid phase *i e* in the bone in favor of the liquid phase or the blood The etiologic relationship between hyperparathyroidism and generalized osteitis fibrosa cystica has been established on a scientific basis The disease has been produced experimentally in animals by the injection of parathormone Patients with cystic bone lesions have recovered after parathyroidectomy The association of characteristic bone lesions with pathologic fractures hypophosphatemia, hypercalcemia and parathyroid tumors has been proven In some instances there is generalized hypertrophy with hyperfunctioning of the parathyroids without actual adenoma formation in the gland Failure to find an adenoma of the parathyroid at operation does not prove definitely that none is present as there are numerous instances on record in which the tumor was found only at autopsy

Blood Chemistry and Urinary Changes In generalized osteitis fibrosa there is hypercalcemia the serum calcium being from 12 0 to 24 0 mg per 100 cc of blood serum (normal is 9 0 to 11 0 mg per 100 cc) The plasma phosphorus is lowered to between 1 0 and 2 0 mg (normal is 2 5 to 3 5 mg per 100 cc) The blood serum phosphatase (Bodansky unit) is elevated from a normal of 2 5 mg to 18 to 20 mg per 100 cc in the presence of demonstrable changes in the bone In Paget s disease and osteomalacia the serum calcium is normal or slightly low With metastatic bone carcinoma and multiple myeloma there may be slight hypercalcemia In active rickets osteomalacia and Paget s disease, the phosphatase (Bodansky) level is raised There is increased output of calcium and phosphorus in the urine sometimes six to eight times the normal amount The metabolism of calcium is increased while that of phosphorus is decreased These factors are elevated in the urine as noted above The balance of calcium and phosphorus shows loss of both from the body the amounts excreted being greater than the intake There is also a disturbance in the normal ratio of the excretion of calcium in the

urine and feces In parathyroidism, there is approximately 70 per cent in the urine and 30 per cent in the feces normally there is 10 to 30 per cent in the former and 70 to 90 per cent in the latter In consequence, stones and calcium phosphate precipitates are found in the renal parenchyma

Pathology Tumors of one or more of the parathyroid glands are found regularly The causal relationship between tumors of the parathyroid and the bone changes in Recklinghausen s tumors of the parathyroid has been appreciated only in recent years For this reason many authors prefer the term hyperparathyroidism Operative removal of the parathyroid tumors leads to healing and proves that the primary cause of the disturbance of the skeleton is actually hypersecretion of parathormone The blood chemistry and urine chemistry in osteitis fibrosis cystica generalisata parallel the findings in experimental hyperparathyroidism There is heightened activity of osteoclasts and of metastatic calcification It has been found that certain symptoms of clinical hyperparathyroidism do not have a counterpart in experimentation An important difference is the presence of extensive new bone formation in osteitis fibrosa cystica generalisata This is probably due to the fact that it is not possible to maintain experimental hyperparathyroidism the same length of time as the duration of the clinical disease hyperparathyroidism, in humans The newly formed bone is immature coarse, fibrillar spongy, and presents all the characteristics seen in rapid bone formation The matrix of the immature bone may show incomplete calcification and portions of the bone remain uncalcified persisting for long periods as osteoid tissue The failure of calcification is assumed to be due to the inhibiting action of the parathyroid hormone on vitamin D In severe advanced cases the compact cortical bone disappears completely and is replaced by a thick layer of spongy bone The trabeculations are thin and the marrow spaces become filled with fibrous tissue The marrow cavity may be markedly narrowed The marrow undergoes metamorphosis to fibrous tissue over extensive areas The lungs, stomach arteries and kidneys are the sites of multiple metastatic calcifications In the kidney there may be extensive damage with manifestations similar to those in Bright s disease In osteitis fibrosa cystica generalisata there develop the so called giant cell tumors or brown nodes Some of the nodes are later replaced by cysts while others heal and disappear by the formation of new bone The cysts are usually the result of hemorrhage in the bone Liquefaction of the blood clot and progressive growth of the primary cyst may lead to the formation of large cavities in bone Solitary giant cell nodes occur in areas which are most commonly exposed to trauma such as the distal epiphysis of the femur the proximal epiphysis of the tibia and the distal end of the radius It is not known why trauma at times leads to the development of the brown node and in other cases to the formation of a primary cyst

Roentgen Manifestations There is marked bone decalcification, first seen as osteoporosis with general or granular mottling Later small cystic areas appear These enlarge to oval or rounded areas of radiance with sharply defined borders and multiple trabeculae the bone cysts or osteoclastomas The large cysts and giant cell tumors expand and thin the cortex Deformities and pathologic fractures occur frequently The changes in the skull are marked The inner and outer tables are largely or completely obliterated Isolated discrete cystic areas occur throughout

the calvaria The bones show marked osteoporosis and diffuse stippling Scattered foci of increased density may alternate with patches of rarefaction giving a tufted or wooly appearance Areas of cystic resorption may expand and produce marked deformations The sinuses appear large The sella and other anatomic landmarks are poorly defined because of the marked decalcification The base of the skull may bulge upward as in Paget's disease The films appear poor and improperly exposed because of the extreme decalcification of the skull bones The mandibles and maxillas show osteolysis and large osteoclastomas The giant cell tumors may cause marked expansion of the bone with a trabeculated appearance of the bone

In the early stages there is generalized osteoporosis with curvatures of the long bones and flattening of the pelvis The vaults of the skull show thickening and local or generalized osteoporosis Cysts and tumors of the bones develop rapidly and may occur in any portion of the skeleton but are found principally in the diaphyses of the long bones The cysts vary greatly in number and size and may be endosteal or subperiosteal in location The cortex may be expanded or merely greatly thinned without expansion The periosteum shows no evidence of proliferation and soft tissue changes are not present The portions of the bones not involved by the cyst formation show marked osteoporosis Pathologic fractures occur in relation to the cysts and produce marked deformities, particularly as non union is common Multiple cysts occur in the maxilla and mandible In the vertebrae there are many small cystic areas and the bodies of the vertebrae may show narrowing The ribs and bones of the pelvis present many cysts of varying size The cysts are trabeculated and non trabeculated Localized overgrowth of bone in the shafts of the long bones produces tumor like formations of irregular spongy bone with a thin cortex

It must be stressed that in only approximately one third of the patients with the disease is there sufficient change in the bone to make roentgen diagnosis possible Most cases are discovered during study for urinary tract calculi While the generalized bone manifestations are of interest, they are not of great assistance in establishing the diagnosis by roentgen methods in many instances The changes which occur in the phalanges of the fingers are pathognomonic These comprise a lace like decalcification of the cortical bone directly beneath the periosteum a sub periosteal resorption of bone This may be shown best in the middle phalanges and is usually accompanied by a resorption of the terminal tufts of the distal phalanges While these changes occur in primary hyperparathyroidism and are considered characteristic of the disease it must be stressed that similar changes may occur in secondary hyperparathyroidism associated with chronic renal insufficiency An important manifestation is the absorption of the lamina dura which represents the cortex of the bone around the teeth This sign is not as reliable as subperiosteal resorption of bone in the phalanges since it may also occur in malacic conditions and in early Paget s disease The lamina is not present in people who are edentulous

A regional or localized form of fibrocystic disease is characterized by involvement limited to the long bones of a single extremity the other bones showing no changes There is central cortical destruction with replacement by fibrous tissue The shaft is not expanded and there is little or no peripheral new bone formation The entire diaphyses of the long bones are involved and pathologic fractures curvature and shorten

ing are common Cysts seldom occur The blood calcium and phosphorus are normal It is considered congenital in origin Sclerosis rather than osteoporosis is the outstanding feature

Differential Diagnosis In differential diagnosis it is necessary to consider metastatic carcinoma, multiple myeloma, osteomalacia lymphoma, syphilis renal rickets xanthomatosis, basophilism, eosinophilic granuloma fibrous dysplasia of bone, and other similar lesions In metastatic carcinoma and multiple myeloma, there is absence of the generalized decalcification and mottling which are found in parathyroidism Osteoporosis is due to a deficiency of the organic matrix of bone, and results from a lack of activity of the osteoblasts It occurs in hypogonadism, particularly the post menopausal state, senility, immobilization, protein insufficiency such as develops in Cushing's disease and starvation Subperiosteal resorption does not occur and there is not the granular decalcification of the skull which is seen in hyperparathyroidism In renal rickets or renal osteodystrophy there are changes which by roentgen methods cannot be distinguished from those of primary hyperparathyroidism due to the fact that renal insufficiency of long standing induces bone changes which are the same as those in hyperparathyroidism This is of extreme importance since operation is often curative in primary hyperparathyroidism but definitely harmful in renal osteodystrophy Primary hyperparathyroidism seldom develops in children Cyst-like lesions in the bone do not occur as frequently in secondary hyperparathyroidism as in the primary form Extensive vascular calcification is frequent in renal osteodystrophy but is rare in primary hyperparathyroidism Fibrous dysplasia of bone is a localized disturbance of growth and development associated with cyst like lesions which superficially resemble those in hyperparathyroidism The areas which suggest cysts in fibrous dysplasia are characterized by replacement of the spongiosa and marrow by fibrous tissue Changes in the phalanges and the lamina dura do not develop in fibrous dysplasia and the skull changes are different In osteomalacia, there is a normal laying down of osteoid with defective calcification of the organic matrix This disease may be the result of poor absorption of calcium in the gastrointestinal tract or resistance to the action of vitamin D It may also ensue after renal tubular disease with excessive excretion of calcium or phosphorus Pseudo fractures frequently occur The subperiosteal resorption of the phalanges is not found in osteomalacia Marked subperiosteal resorption of bone in regions adjacent to the joints results in an appearance similar to that in rheumatoid arthritis Similar changes may occur in the phalanges of the feet the medial aspect of the upper third of the tibia the outer end of the clavicle and the margins of the distal end of the ulna After hyperparathyroidism has been corrected by operation the subperiosteal bone shows rapid regeneration and the cortex again assumes a normal appearance This is also true of the tufts of the phalanges

In osteomalacia rickets carcinomatosis multiple myeloma and pregnancy hypertrophy of the parathyroid glands may occur However none of these conditions presents roentgen evidence of secondary hyperparathyroidism This is probably due to the fact that in hypertrophy of the parathyroid gland there is a well regulated compensatory mechanism The type of parathyroid response which occurs in renal insufficiency suggests that it may be truly toxic and not compensatory in origin Subperiosteal resorption of bone is of importance and is a great aid in the

diagnosis of primary hyperparathyroidism and renal osteodystrophy It does not occur in other conditions and is pathognomonic of these diseases Studies of a series of cases of osteomalacia reveal no evidence of subperiosteal resorption despite the fact that there is marked decalcification There never was found any suggestion of the lace like appearance of the subperiosteal bone which is characteristically seen in primary hyperparathyroidism and renal osteodystrophy Subperiosteal resorption does not occur in postmenopausal and senile osteoporosis or Cushing s disease Cases of hypervitaminosis D present extensive decalcification of bone and periarticular calcinosis but no evidence of subperiosteal resorption of bone

Treatment Since hyperparathyroidism is associated with hypertrophy or hyperplasia of one or more parathyroid bodies the treatment of choice is parathyroidectomy Excellent results have been obtained with this procedure However it is difficult to find the parathyroid bodies They may be encountered anywhere from the pharynx to the mediastinum If not removed at the first operation, the formation of scar tissue makes subsequent operation more difficult In many instances there is such a marked degree of cachexia that surgical intervention is not feasible In certain instances irradiation produces results which compare favorably with those of surgery

Prognosis The course of the disease varies widely Most frequently there is rapid increase in the extent of involvement The patient grows progressively weaker and death usually results from intercurrent infection Cases of spontaneous regression with healing and disappearance of the lesions are on record Operative removal of parathyroid neoplasms may produce improvement although relapses occur in many instances

Secondary Hyperparathyroidism

Hypertrophy and consequent hypersecretion of the parathyroid are known to occur in various experimental and clinical circumstances The repeated injection of phosphates results in one form of secondary hyperparathyroidism The mechanism apparently is in accord with the action of the parathyroids upon the kidneys there being a decrease in reabsorption with resultant increase in excretion of phosphorus The increased elimination of phosphate because of hyperactivity of the parathyroid glands is a direct response to injection of phosphates Calcium deficiency leads to secondary parathyroid hypertrophy This is explained as follows The calcium blood level is decreased and the organism attempts to restore the normal level by hyperactivity of the parathyroid This results in mobilization of calcium from the skeleton The end result of the calcium deficiency is a generalized osteoporosis Calcium deficiency is rarely if ever the result of a lack of calcium in the diet the deficiency rather being due to failure of the organism to absorb calcium The absorption of calcium is intimately linked with the absorption of fat Impairment of fat absorption because of lack of bile as in fistula of the common bile duct or disease of the intestinal tract as in celiac disease results in calcium deficiency Secondary hyperparathyroidism also occurs in vitamin D deficiency

Experimentally hyperfunction of the parathyroid gland is simulated by the injection of parathyroid hormone Within a few minutes after the injection of the parathyroid hormone the amount of phosphorus in the

urine shows an appreciable elevation Normally, phosphorus is filtered in the glomerulus of the kidney and undergoes reabsorption to a considerable extent in the convoluted tubules The resorption of phosphorus is depressed or prevented by the administration of parathormone In consequence of the hypersecretion of phosphorus, the phosphorus blood level is depressed Histologic examination of the bones of an animal twelve hours after injection of the hormone reveals a marked increase in the number of osteoclasts and a decrease in the osteoblasts Osteoporosis, replacement of the bone marrow by young connective tissue with many giant cells and resultant fibrosis of the bone marrow ensue The giant cells in the tissues of the widened marrow spaces appear to be osteoblasts which have survived the destruction of bone trabeculae After the onset of the changes in the bone study of the blood reveals an increase in the calcium level Not all of the bones or all parts of the bone are affected to the same extent The rapidly growing bones or areas of bone are more severely affected This phenomenon can be appreciated more clearly if it is borne in mind that a more rapid rate of growth indicates *not only* more rapid apposition but also a higher rate of reconstruction and therefore more rapid resorption The reaction of the bone after the injection of parathormone constitutes an exaggeration of normal osteoclastic activity

Repeated injections of large doses of parathormone during a long period of time result in gradual decrease of the effect of the hormone Eventually complete healing of the primary damage takes place It is believed that this is due to the fact that there is an elaboration of an antihormone which neutralizes the hormone which has been injected The administration of small amounts of hormone during long periods of time stimulates the formation of large amounts of antihormone and after a short period of osteoclastic hyperactivity there ensues increased osteoblastic activity with resultant osteosclerosis Additional doses of the hormone result in the establishment of a normal balance of apposition and resorption of bone The hyperproduction of bone is explained on the basis that the first injections of small doses of the hormones result in excessive production of antihormone which neutralizes the hormone produced by the glands A secondary hypoparathyroidism ensues with resultant hyperproduction of bone

Latent Hyperparathyroidism In cases of renal insufficiency there develop localized and in some instances minute bone changes These manifestations have been noted in the vertebræ the maxilla, and the mandible The localization of such foci of osteitis fibrosa cystica generalisata is not a mere matter of chance The changes occur in areas of the jaw which have suffered a mechanical or an infectious injury such as a bony scar after extraction of the tooth in the neighborhood of infected roots, and also in the region of an inflammatory polyp of the maxillary sinus This indicates that moderate renal insufficiency may create a state of latent hyperparathyroidism which is manifested by increased lability of the skeleton The resultant changes are the response of the sensitized skeleton to a localized injury On microscopic examination the typical manifestations of osteitis fibrosa cystica localisata are found, there being increased resorption of bone, fibrous bone marrow and the formation of immature bone which in some instances does not calcify On roentgen study the foci of osteitis fibrosa are manifested as defects in the bone because the immature bone is much more radiolucent than

mature bone despite the fact that it is partially calcified The difference in density is due to the fact that the immature bone contains more cells and less intercellular substance and therefore less calcium than is the case with mature bone

Hypoparathyroidism

Hypoparathyroidism has an effect opposite to that of hyperparathyroidism Bone resorption is decreased The kidneys reabsorb more phosphorus and the blood level of phosphorus rises The organism disposes of the surplus phosphorus by combining it with calcium and depositing it in the form of newly produced bone or as metastatic calcification The calcium required in the process of bone formation is derived from the blood consequently the calcium blood level falls below normal There is also overexcitability of the nerve endings particularly the motor nerves Bone resorption becomes greatly diminished The combination of increased apposition and decreased resorption of bone leads to progressive osteosclerosis In humans there are three types of hypoparathyroidism (1) postoperative (2) neonatal and (3) idiopathic The most striking manifestation of hypoparathyroidism is tetany Tetany is caused by deficiency of calcium in the blood serum The normal action of calcium is to diminish the irritability of the nerve endings In the rare cases in which the parathyroid glands are removed in a goiter operation the resultant hypoparathyroidism is in all respects analogous to that which occurs after experimental parathyroidectomy The second type is found in infants whose mothers had had a compensatory hyperparathyroidism during pregnancy This may be caused by an extreme calcium deficiency in the diet The fetus is provided with the necessary amount of calcium at the cost of the blood calcium of the mother and the maternal parathyroids hypertrophy to mobilize calcium from the skeleton In consequence of the maternal hyperparathyroidism the fetus obtains directly from the mother more parathyroid hormone than it would under *normal conditions and the development of its own parathyroids is arrested* Soon after birth this lack of parathyroid function is manifested as tetany The tetany disappears after the delayed but complete development of the parathyroid A third type is idiopathic hypoparathyroidism Marked osteosclerosis ensues and is demonstrable by roentgen examination There is an apparent contradiction in the fact that the newly formed bone tends to fail to calcify and that simultaneously there develop multiple metastatic calcifications The assumption that a specific action of the parathyroid hormone inhibits the calcifying action of vitamin D appears to explain this phenomenon Calcification of osteoid tissue is associated with local changes in the intercellular substance and in this mechanism the osteoblasts apparently play an active role When these changes which are normally stimulated by the action of vitamin D are inhibited there is persistence of osteoid tissue

SYMMETRIC CEREBRAL CALCIFICATION OF THE BASAL GANGLIA ASSOCIATED WITH PARATHYROID INSUFFICIENCY In 1938 Eaton Camp and Love reported a characteristic roentgenographic symmetric cerebral calcification most marked in the basal ganglia In this condition the degree of calcification varies widely according to the stage of the disease Early, deposits of calcium are small and irregular and are not demonstrable on the roentgenogram In the marked cases the distribution is more generalized, calcifications occurring in regions other than the basal ganglia

and at times coalescing into a more or less homogeneous mass. Roentgen study reveals shadows which are bilateral and symmetrical and are apt to be confused with those due to brain tumor, although pneumoencephalography may be necessary for final diagnosis. Other conditions which produce abnormal densities are abscess, torulosis, calcific deposits in the walls of the ventricles, calcified intraventricular hemorrhages, tuberous sclerosis, and toxoplasmosis. Calcifications of the choroid plexus lie in the genu of the lateral ventricle and are well posterior to those in the basal ganglia. In symmetric cerebral calcification of the basal ganglia, the changes apparently are due to colloid deposition in and around the finer cerebral blood vessels with ensuing calcium deposits. The patients may exhibit tetany, mental deterioration, and convulsive seizures.

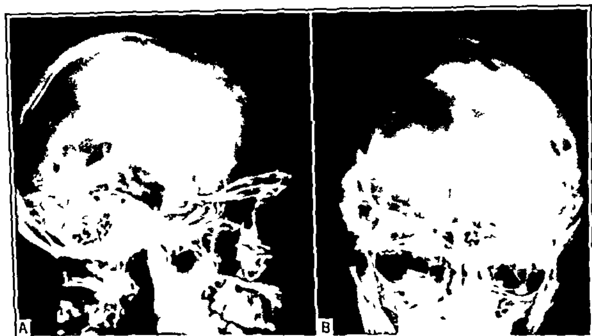


Fig. 191. Symmetric Cerebral Calcification of the Basal Ganglia. *A*, Lateral view. There is irregular mottled calcification in the temporoparietal regions. *B*, Anteroposterior view. The calcific deposits are symmetrical, bilateral and irregularly mottled. The distribution of the calcifications in the sagittal projection indicates that the depositions are in the basal ganglia.

Eaton, Camp, and Love indicate that there is an association with spontaneous parathyroid insufficiency. They also report a case in which symmetrical cerebral calcification developed in a patient in whom parathyroid insufficiency became manifest following parathyroidectomy. This supports their opinion that the cerebral changes are secondary to the metabolic disturbances resulting from insufficient parathyroid secretion and that the cerebral changes are not the primary cause of the disturbance.

A case seen by us was first observed about three years ago. The calcific deposits were noted during a routine examination of the sinuses made because of a "head cold" with sinusitis. The patient was a man, aged thirty-seven years, who had no previous complaints and was otherwise apparently in good health. The roentgenograms revealed irregular mottled calcifications in the parietal and temporal regions which suggested in their distribution the outlines of the lateral ventricles. In the sagittal views, symmetrically bilateral calcific deposits in the form of curved lines

lying to either side of the midline were demonstrable Encephalography carried out at another hospital definitely demonstrated that the calcific deposits were outside the ventricles Re examination of the patient two years later showed a slight increase in the size of the calcific densities The patient became extremely concerned when he learned of the calcifications and complained of marked weakness, lassitude and frontal headaches All clinical and neurologic examinations have shown no abnormality and the eye grounds have been clear No abnormal calcific deposits have been detected roentgenographically elsewhere in the body except for slight hypertrophic changes in the lower spine, although these are not greater than might be found in an individual of this age

Pseudohypoparathyroidism Seabright Bantam Syndrome

Pseudohypoparathyroidism is a rare disease only a small number of cases having been recorded in the literature The clinical picture is characteristic There is tetany with convulsions carpopedal spasm laryngeal spasm and positive Chvostek s and Trousseau's sign There are characteristic alterations in the configuration of the body which suggest but are not typical of achondroplasia with short stature obesity sturdy build, rounded features and irregularities in length of the metacarpals and the metatarsals Areas of calcification are present in the soft tissues There is decalcification of the teeth Mental retardation is a prominent feature The characteristic blood findings comprise a reduction in the serum calcium and elevation of the serum inorganic phosphorus the alkaline phosphatase remaining unchanged The patients do not respond to the injection of parathyroid extracts with an increase in the urinary output of phosphorus and an elevation of the serum calcium as do normal individuals and patients with true hypoparathyroidism Because of this, Albright decided that the metabolic defect is an insensitivity to the receptors of parathormone

The name *Seabright Bantam syndrome* has been applied to one form of pseudohypoparathyroidism because of the analogy to the condition observed in the Seabright Bantam roosters These roosters have female tail feathers presumably because of failure to respond to male sex hormones The condition in humans is believed to be a failure of the end organ of the kidney to respond to parathormone despite the fact that there is adequate production of the hormone Treatment consists of the administration of vitamin D in high doses and the oral administration of calcium

On roentgen examination there is generalized thickening of the skull The bones of the forearm are broad and gnarled The fibulas are bowed inward with increased subperiosteal density There are deformities of the metacarpals, the distal ends showing sharply demarcated areas of decreased density as in chondrodystrophy The metacarpals are short and broad although in some instances the phalanges may be long The shortening of the metacarpals affects particularly the first fourth and fifth metacarpals These bones are late in developing endochondral bone and slow in the formation of the secondary centers of ossification There may also be increased length of the metatarsals The phalanges of the feet are distorted as in chondrodystrophy The spine shows osteoporosis,

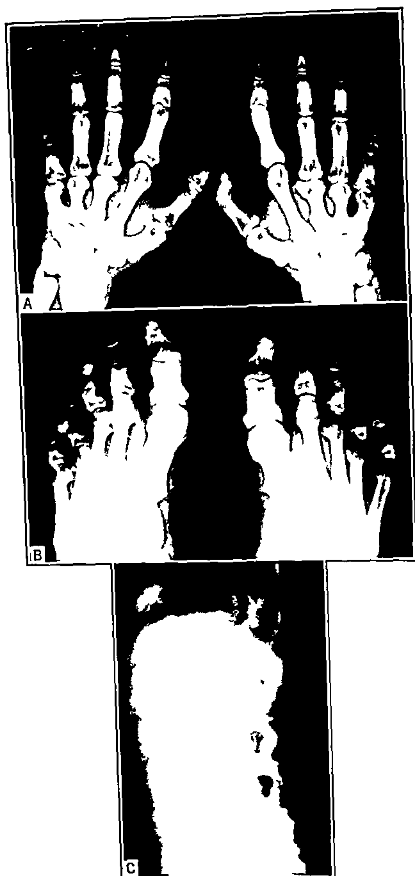


Fig 197 Pseudohypoparathyroidism Seabright Bantam Syndrome *A* Hands The metacarpals with the exception of the second are markedly shortened with broad blunt extremities The proximal interphalangeal joints of the index fingers are irregular in outline The bones of the fingers are osteoporotic *B* Feet The changes in the bones of the feet are analagous to those in the hands *C* Dorsolumbar Spine The superior and inferior surfaces of the vertebral bodies show concave defects irregularity of outline and slight eburnation The bones are porotic

biconcave defects in the superior and inferior surfaces of the bodies of the vertebrae, and irregularity of outline of the vertebrae. The changes may be due to more than a deficiency of the end organ response and are now considered as representing a developmental anomaly on a genetic basis although postconceptional and environmental factors also appear *to play a part. There are multiple, widely scattered subcutaneous calci*fications. Para-articular and basal ganglionic calcifications may occur.

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VITAMIN DISTURBANCES—INFLUENCE OF VITAMINS

In order to safeguard the function, development, and growth of the body it is essential that the diet contain proteins, carbohydrates, fats certain minerals and small amounts of elements termed vitamins which are distributed by the blood stream to the organs of the body and act in specific manners on the various tissues The vitamins are analogous to the hormones in their modes of action and have been described as extrinsic hormones The nomenclature of the vitamins is complex and still in a state of flux One system designates the various members of this group by the letters A, B C, etc , the order being according to the time of their discovery A common practice is to utilize the appropriate chemical terms particularly in view of the fact that a single name had been applied to an entire group rather than to a single member of the family Thus vitamin C is known as ascorbic acid Vitamin B has been subdivided into B_1 B B_3 etc each of these being designated by a chemical name such as thiamin and riboflavin Another method of nomenclature is to designate the vitamin by the lesion with which it is associated, for example vitamin C is termed the antiscorbutic vitamin since lack of this factor in the diet results in scorbutus or scurvy The vitamins A, B, C and D exert definite influences on the skeleton, the effects of the other vitamins on the skeleton being as yet unknown

DEFICIENCY OF VITAMIN A

Vitamin A is contained in eggs milk and liver Provitamin A or carotene, which is converted in the body to vitamin A, is found in yellow and green vegetables Vitamin A deficiency may be produced by malnutrition diseases of the gastrointestinal tract which result in lack of absorption and disease of the liver Vitamin A is stored in great quantities in the liver and liver damage prevents mobilization of vitamin A The principal and most characteristic manifestations of avitaminosis A comprise changes in the epithelium Highly differentiated epithelium undergoes atrophy and is replaced by keratinized squamous epithelium derived from the proliferating basal cell layer These changes occur in the epithelium of the salivary glands, nasal cavities trachea bronchi genitourinary tract cornea and the glands of the conjunctiva and lids An important characteristic of vitamin A deficiency is night blindness due to failure of the retina to produce rhodopsin the visual purple Alterations in the skeleton are variable Endochondral bone shows retardation and subsequently total cessation of growth While the alterations at the junction of the epiphysis and diaphysis are not specific there is evidence of interruption of longitudinal growth The rows of cartilage cells and bony trabeculae are shortened indicative of a decrease in proliferation of cartilage and also of its resorption and replacement by bone In animal experimentation avitaminosis A is found to result in retardation of formation of new bone in the alveolar process with consequent delay in eruption of the teeth There is delay in endochondral bone formation and marked inhibition of tubulation The progression of appositional bone formation is normal In dogs there is retardation of bone growth malposition of the teeth and malformation of the roots of the teeth Neurologic changes formerly were considered a primary effect of deficiency of vitamin A

These manifestations have been proven to be secondary to disturbances in the growth of the cranial bones and the vertebrae, the nerve degenerations and resultant paralyses being mechanical in origin There is disproportion between the size of the skull and the vertebral column It is not known whether the disproportionate size of the brain and spinal cord is the result of cessation of skeletal growth while growth of the brain continues or whether there is encroachment of the bones of the skull upon the space normally occupied by the brain due to overgrowth of the bones There are evidences of overgrowth of periosteal bone in the labyrinthine regions of the skull Sutural growth is inhibited Wolbach and Sicher stress the fact that the tissue changes which contribute to bone growth suffer in progressive order Proliferation, degeneration, calcification and resorption of cartilage are the first to cease Subsequently there is cessation of proliferation of sutural connective tissue the sole factor in sutural growth The last to be affected is the simple apposition of bone as for example on periosteal surfaces The difficulties in explaining the genesis of the changes in vitamin A deficiency are complicated by renal damage, there being calcification of the epithelium of the convoluted tubules and cloudy swelling of the cells of the collecting tubules in addition to cornification of the epithelium of the renal pelvis The osteoporosis in vitamin A deficiency can be explained on the basis of pathologic changes in bone of renal origin

HYPERVITAMINOSIS A

Joseph in 1944 was the first to report that excessive intake of vitamin A may produce toxic effects in humans The condition is often not suspected by the parents or clinician as there is a lack of awareness of the potential dangers of overfeeding There is a latent period of six to eighteen months between the beginning of the excessive intake of the vitamin and the onset of the toxic manifestations hence the etiologic relationship between the ingestion of the vitamin and the development of the symptoms may be overlooked Prolonged and continuous administration of large amounts of vitamin A appears necessary to cause harmful effects In the previously reported cases the amount of vitamin A ingested has varied from 60 000 to 600 000 units per day The use of large doses of vitamin A in the treatment of skin diseases is potentially dangerous as instances of intoxication have developed in this manner The primary organ concerned with vitamin A metabolism is the liver Its capacity to store this vitamin is enormous and it can maintain 95 per cent of the total vitamin A reserve The ingestion of amounts which overwhelm the capacity of the liver to remove vitamin A from the blood becomes harmful Clinically all cases of the disease have disclosed hepatomegaly although no evidence of hepatic dysfunction is demonstrable

The clinical manifestations consist of anorexia irritability pruritus skin rash tender swellings subcutaneously pain over the extremities, cracked dry lips and sparse coarse hair In infants and children there is fretfulness and the patient resists being touched or moved Low grade fever and hepatomegaly may occur The spleen is not enlarged There is an abnormally high vitamin A level in the blood serum and the blood lipids may be elevated The serum phosphatase is not increased as a rule In some instances there is an abnormal craving for butter, cream

and other foods which contain large amounts of vitamin A After withdrawal of the offending vitamin, the symptoms regress rapidly Within about four weeks, the tenderness and swelling of the extremities disappear The changes in the bones and the elevated vitamin A level in the blood may persist for a considerable period after the patient appears to have recovered clinically The prognosis in cases in which the excessive intake of vitamin A has been eliminated is excellent Acute poisoning due to the ingestion of a single large dose of the vitamin is rare but has been reported in children

Roentgen Manifestations Roentgen study reveals extensive hyperostoses with new bone formation involving the shafts of the long bones The newly formed bone may be lamellated The process affects two or more bones The hyperostoses present a shell like appearance with a zone of increased radiance between the original cortex and the new bone The ulnas are affected in practically every instance In the clavicle the involvement may initially be confined to the distal segment alone, subsequently the entire bone is affected The changes in the ribs are frequently limited to the lateral axillary portions The femurs show bony overgrowth along the upper half of the shaft The involvement of the fibulas is lateral or generalized Elongation and overtubulation of the long bones may occur with an early excessive rate of endochondral bone formation which results in premature closure of the epiphyses Changes in the skull are less common and consist of periosteal new bone formation The new bone is less dense than the normal bone of the skull It is homogeneous in character and is similar in appearance to the hyperostoses in the long bones The cranial hyperostoses may involve the occiput and in this instance there may be coarctation and asymmetry of the foramen magnum After withdrawal of the vitamin the bone changes regress Within one month the hyperostoses of the long bones are distinctly less dense and smaller The changes in the skull persist longer than in the case of the long bones The bony proliferations do not clear as rapidly as the clinical manifestations and may persist for several months

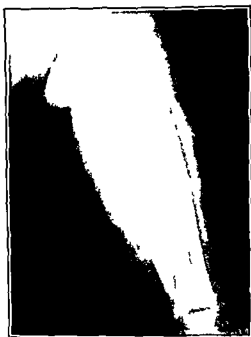


FIG 193 Hypervitaminosis A There is marked periostitis of the fibula with extensive new bone formation involving the entire shaft of the bone Similar changes were present in the ulna clavicle and ribs

Differential Diagnosis Infantile cortical hyperostosis appears during the first four months of life while hypervitaminosis A has not been recognized before twelve months of age The face and jaws are involved in infantile cortical hyperostosis but not in hypervitaminosis A The metatarsal bones are frequently affected in hypervitaminosis A but only rarely in infantile cortical hyperostosis The blood vitamin A is increased in hypervitaminosis A Fever is the rule in infantile cortical hyperostosis

but is absent in hypervitaminosis A. Recovery from hypervitaminosis A is rapid after cessation of ingestion of excessive doses of the vitamin while recovery in infantile cortical hyperostosis is slow. Lesions of the scapula are unknown in hypervitaminosis A but are common in infantile cortical hyperostosis.

VITAMIN B DEFICIENCY

The important vitamins which comprise the vitamin B complex are B_1 or thiamin, the beriberi preventing vitamin; B or riboflavin; the nicotinic or pellagra preventing factor; the filtrate factor which consists of pantothenic acid; and other less well known elements. The B complex vitamins are found in abundance in lean meat, particularly pork, liver, heart, and other organs, in various vegetables, especially potatoes and carrots, and in rice polishings. The effects of these vitamins on the human skeleton is not clearly understood. A deficiency of riboflavin in the diet of female rats is responsible for congenital skeletal malformations in the offspring. It appears that maternal nutritional deficiency may result in arrest of development of the embryo with resultant congenital deformities of the offspring. Death usually does not ensue. The malformations which have been reported comprise shortening or absence of the tibia, fibula, radius, ulna, and bones of the feet, shortening of the mandible, fusions of the ribs, and various deformities of the sternum, maxilla, clavicle and scapula. Microscopic studies show that the shortening of the long bones occurs in the cartilaginous stage and ossification is retarded. Deformities of the cartilaginous model lead to abnormalities of shape and structure of the bone.

Studies in dogs to determine the consequences of deficiency of the filtrate factor and nicotinic acid show that in nicotinic acid deficiency there develops an inflammation of the gingival tissues. Deficiency of the filtrate factor leads to osteoporosis. The most extensive osteoporosis occurs in the upper jaw. Deficiency of both nicotinic acid and the filtrate factor results in a combination of inflammatory and osteoporotic changes.

VITAMIN C DEFICIENCY—SCURVY

The first accurate and comprehensive description of the cause, symptoms and pathology of scurvy in adults was by Lind in 1747. The recognition of infantile scurvy, originally called acute rickets, was made by Barlow in the latter part of the nineteenth century. The disease had become increasingly prevalent at that time because of the use of artificial methods of infant feeding. Barlow stressed the frequency and severity of the subperiosteal hemorrhages which occurred in the long bones and less commonly also in the flat bones such as the scapula, the cranial vault and the orbits. He considered the subperiosteal hemorrhages analogous to the hemorrhages which occurred in the gums and the skin in adult scurvy and was of the opinion that they caused a disturbance in the growth process at the ends of the long bones which resulted in epiphyseal separation and displacement. The early bone changes in infants were studied by means of the recently discovered roentgen rays by Frankel. His name has been applied to the metaphyseal line of increased density which he stressed as the important sign of scurvy. Wimberger described the signet ring appearance of the epiphyses which has since been referred

to by his name It has only gradually become evident that the subperiosteal hemorrhages are a late manifestation of scurvy and that the earliest bone changes occur in the metaphyses The sex distribution in scurvy is about equal The age incidence reveals a pronounced limitation of the disease to the latter half of the first year of life In a large series of cases studied by Evans no cases were observed before the seventh month and 90 per cent fell in the age period from seven to thirteen months inclusive The most advanced lesions were usually found in the knees, the shoulders, ankles wrists, hips and elbows being less frequently affected

The antiscorbutic vitamin C is contained in high concentration in citrus fruits, green peppers, leafy vegetables, and walnuts before they ripen Ascorbic acid has been synthesized Deficiency of vitamin C



FIG 194 Scurvy The epiphyses are granular in appearance and show increased radiance except for a peripheral rim of increased density There is generalized osteoporosis The epiphysis of the lower end of the left femur is displaced laterally

presents a characteristic clinical picture known as scurvy or scorbutus This was one of the most dreaded dangers of long voyages on sailing ships during prolonged military sieges and on arctic and antarctic expeditions during which people had to live on preserved or insufficient foods for long periods of time While the disease is relatively uncommon today it still occurs in infants fed artificial diets, in adults on an ill-balanced diet in patients suffering from alcoholism and in others on a near starvation diet The disease presents a characteristic clinical picture There is extensive hemorrhage in the muscles of the extremities particularly the extensors with resultant pseudoparalysis and subperiosteal hemorrhages especially in the vicinity of the joints Swelling and bleeding of the gums and loosening or loss of the teeth are common If wounds develop they fail to heal In infancy and childhood the patient assumes a characteristic position in the advanced cases The knees,

hips shoulders and elbows are slightly bent and are abducted and rotated laterally because pain in the muscles and joints forces the patient to assume the position in which all or most of the parts are relaxed Separation of the epiphyses occurs in children The separation occurs most commonly in the lower epiphysis of the femur and tibia and the upper epiphysis of the humerus It is believed that the separations are not spontaneous but are the result of slight trauma

Roentgen Manifestations In scurvy there is a marked degree of generalized atrophy of the bones thickening widening and increased density of the epiphyseal plates and a zone of increased density about the ossification centers There is a zone of rarefaction between the thickened epiphyseal plate and the shaft of the long bones Lateral spurs form at the ends of the diaphyses and there are clefts and crevices in the angles between the epiphyseal plate and the cortex The cortex of the bones is markedly thinned An important manifestation is extensive subperiosteal hemorrhage with fractures and separations of the epiphyseal plate and displacement of the epiphysis This is most commonly seen in the knees wrists and ankles but may also occur in the flat bones such as the scapula the cranial vault and the orbits The earliest changes comprise a ground glass type of demineralization of the bone structure and a signet-ring appearance of the epiphyses The signet ring appearance is less constant being found in about 75 per cent of the cases It may be absent in cases complicated by rickets The submetaphyseal notch which has been stressed by many authors as the earliest pathognomonic sign of scurvy is almost constantly present but may be masked by co existent rickets The metaphyses may be fragmented or separated and actual displacement of the entire metaphysis or a large segment carrying with it the epiphysis may occur Marked elevation of the periosteum occurs along the diaphyses of the long bones due to subperiosteal hemorrhages The periosteal shadows are of two types The first form appears as a narrow triangular shadow with the base at the metaphysis and extending for a short distance along the shaft This develops in association with fragmentation of the metaphysis and slight lateral displacement of a marginal fragment at the lateral or medial aspect of the bone The second type of periosteal change is more extensive and is manifested by a club shaped appearance extending along practically the entire length of the shaft It is almost constantly associated with displacement of the epiphyses The subperiosteal hemorrhage is demonstrable by roentgen methods only when calcification begins In the presence of a large subperiosteal hemorrhage the displacement of the epiphyses may not be demonstrable unless the roentgen exposures are made in various planes The most common site of epiphyseal displacement is the lower end of the femur The displacement is laterally hence if the roentgen examination is made with the child on the back and the legs in external rotation and abduction lateral views of the knees will result and the displacement of the epiphyses may not be demonstrable Epiphyseal displacement may not be demonstrable because it is not present at the moment the x-ray study is made It is well known that in epiphyseal displacement the epiphyses are mobile and the epiphyses may by chance be in normal position despite the fact that there recently had been marked displacement and a large subperiosteal hemorrhage

In children the significant roentgen manifestations comprise the following triad (1) the shaft of the long bone has a ground



Fig 195 Scurvy The flaring of the anterior aspects of the ribs is similar to that in the so-called rachitic rosary and often results in an erroneous diagnosis of rickets



Fig 196 Scurvy There is a dense shell of subperiosteal bone about the subperiosteal hematoma The infant is six months old and has scurvy which is undergoing healing

glass appearance due to osteoporosis, (2) the zone of preparatory calcification of the epiphyseal cartilage is widened and is demonstrable by roentgen examination as an accentuated, broadened line, (3) the region of the metaphysis bordering the epiphyseal line comprises a zone of rarefaction. In untreated cases, calcification ensues in layers parallel to the shaft in areas of hemorrhage beneath the periosteum. With healing, there is thickening of the cortex, increased prominence of the spongiosa, disappearance of the zone of rarefaction, and increase in width of the dense ring about the ossification centers. If subperiosteal hemorrhage had occurred, deposition of calcium ensues after administration of vitamin C with resultant organization and calcification of the hematoma. As the patient grows, the thickened epiphyseal plate is buried in the shaft and appears as a dense white line extending transversely across the bone. The cuff of calcified hematoma is gradually absorbed and the epiphyseal fractures and displacements heal without orthopedic treatment.

Pathology. The histologic changes are characteristic. The preparatory calcification of cartilage is not impaired and the zone of calcification is widened. The apposition of bony trabeculae is greatly disturbed. The cells of the primary marrow produce a homogeneous bone-like substance which is of low differentiation and inferior quality and may form considerable masses at the epiphyseal ends of the metaphyses. The compact bone is often replaced by spongiosa. The junction between bone and cartilage is weakened because of the presence of osteoporosis and lack of interdigitation which normally develops between the regular spicules of cartilage and the trabeculae of bone. Fractures in this portion of the bone occur frequently at the junction of the diaphysis and epiphysis. Bone growth may cease altogether in the acute or the later stages of the disease. The osteoblastic activity is greatly restricted and the amount of bone present in severe cases is reduced markedly. Healing of the weakened bone progresses rapidly by the production of large amounts of densely arranged spongy bone which develop on the periosteal surface of the bone. An important manifestation of scurvy is loosening of the teeth. It is not clearly understood whether the teeth loosen because of damage to the fibroblasts of the peridental membrane or as the result of secondary inflammatory processes involving the gingiva. The hemorrhages have been considered as being due to an increase in the permeability of the capillary walls caused by deterioration of the intercellular cementing substance or the connective tissues around the capillaries, more probably the latter. Similar changes can account for the intramuscular and subperiosteal hemorrhages which usually occur in areas which normally are adapted to mechanical stress by specially organized connective tissue, fascia, muscles or the Sharpey's fibers of the periosteum.

Differential Diagnosis. Rickets is particularly apt to cause confusion in diagnosis and it is difficult to distinguish minor degrees of rickets and scurvy when the two diseases occur simultaneously. The changes characteristic of scurvy may be present in the metaphyses of one bone and those of rickets in another. The rachitic manifestations are apt to be most marked in the distal ulnar metaphyses while the distal radial metaphyses seem to be more prone to the development of the lesions of scurvy. In the case of the knee, the metaphysis is wider and presents a more frayed and irregular appearance with diminished density than when scurvy alone is present. Also the submetaphyseal notch occurs at a greater distance from the epiphyseal line and is less sharply defined. The periosteal mani

festations may afford a distinguishing feature In scurvy the periosteal shadows originate in the metaphyseal areas, are usually widest there and tend to be of uniform density and sharply defined The periosteal shadows of rickets occur in the region of the middle third of the shaft, are frequently laminated, and are most marked in the first six months of life, being much less frequent in the age period when scurvy is apt to occur During infancy the bones frequently become involved by infections with resultant periosteal and osseous lesions The common etiologic factors are syphilis tuberculosis and pyogenic osteomyelitis Clinically, scurvy is easily mistaken for osteomyelitis and not infrequently the periosteal proliferation and involucrum of osteomyelitis have been misdiagnosed as ossifying subperiosteal hemorrhage of scorbutic origin In osteomyelitis during infancy, there may be very extensive irregular periosteal proliferation Ossifying periostitis may be accompanied by displacement of the epiphyses The irregular density and contour of the periosteal shadow are different from that which occur in scurvy The localized destructive lesions in the bone in association with similar lesions in other areas and the absence of changes resembling scurvy in the metaphyses usually establish the diagnosis

Traumatic displacement of an epiphysis results in separation of the periosteum at the end of the shaft with subperiosteal hemorrhage which is in many respects similar to that in scurvy Epiphyseal displacements in infancy are usually the result of trauma These lesions are most common in the proximal humeral and femoral epiphyses and the lower end of the femur The hemorrhage is not as profuse or extensive as in scurvy There is little or no disturbance of the subsequent growth of the bone and no resultant deformity or disability Neoplastic infiltration especially in leukemia and neuroblastoma may produce a picture very similar to that in scurvy There is increased density of the metaphyseal line with submetaphyseal rarefaction in leukemia In scurvy, the line is sharper and denser and offers a greater contrast to the markedly demineralized bone of the diaphysis The submetaphyseal notch is recognizable as a fracture rather than a demineralization as usually occurs in leukemia Subperiosteal leukemic deposits usually involve the mid portions of the shafts of the long bones rather than the metaphyseal areas and there is frequently a recognizable infiltration or destruction of the underlying bone

VITAMIN D

The Action of Vitamin D The ordinary mixed diet is poor in vitamin D Usually however, it contains sufficient amounts of pro vitamin D, cholesterol and ergosterol which change into vitamin D on irradiation with ultra violet rays Cholesterol is stored *in* the skin and is transformed into vitamin D Vitamin D is also obtained by the irradiation of food such as milk The principal sources of vitamin D are egg yolk and dairy products The chief sources of cholesterol are fats The liver stores vitamin D Fish liver particularly of the cod and the halibut contains large amounts of vitamin D Vitamin D deficiency is caused mainly by failure of the body to transform cholesterol to vitamin D because of lack of exposure to sunlight Therapy comprises irradiation with artificial or natural ultra violet light and feeding of vitamin D in fish liver oil or as irradiated cholesterol or ergosterol

The principal function of vitamin D is the regulation of the calcium and phosphorus metabolism by maintenance of the absolute level and physiologic ratio of these two minerals An explanation of the action of vitamin D must take into consideration a series of facts which have been established as the result of studies of clinical and experimental vitamin D deficiency (1) The absorption of calcium and secondarily of phosphorus from the gastrointestinal tract is diminished, (2) The calcium blood level remains normal The phosphorus level is generally depressed (3) The excretion of phosphorus in the urine is increased (4) The parathyroid glands are enlarged, (5) The normal calcification of osteoid and cartilage does not take place

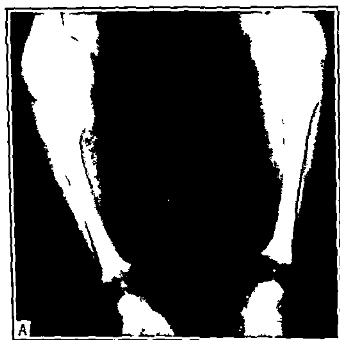
It is believed that vitamin D acts directly to further the absorption of calcium from the intestinal tract and indirectly to effect the reabsorption of phosphorus in the kidneys by depressing the function of the parathyroid glands The lowering of calcium absorption depresses secondarily the absorption of phosphorus because the calcium which remains in the lumen of the intestinal tract combines chemically with the phosphorus with the resultant formation of insoluble calcium phosphates The hyperactivity of the parathyroids which are free of the inhibitory influence of vitamin D, increases the phosphorus deficiency Under the influence of the hypersecretion and later the hypertrophy of the parathyroids, the reabsorption of phosphorus in the renal tubules becomes diminished and in consequence the secretion of phosphorus in the urine is increased A definite fall in the phosphorus blood level indicates that the organism is depleted of phosphorus The level of calcium in the blood in vitamin D deficiency remains practically normal because the absorbed calcium is retained almost entirely in the blood with failure of calcification of cartilage and osteoid tissue It has been stated that vitamin D exerts a direct influence on the calcification of bone and osteoid tissue Observations in cases of primary and secondary hyperparathyroidism indicate that this action is hindered by hyperfunction of the parathyroids In consequence this phase of vitamin D activity is concerned with the changes of osteoid tissue to a cartilaginous matrix which makes these substances calcifiable

The disturbances of calcification in vitamin D deficiency create different pathologic pictures according to the period of life at the time the disease is acquired If the vitamin D deficiency occurs during the period of rapid growth the resultant pathologic change is severe and the clinical picture is that of rickets In the adult the consequences of vitamin D deficiency create various pathologic pictures according to the period of life at the time the disease is acquired The changes in the adult are usually those of osteomalacia and develop only after long periods of time

VITAMIN D DEFICIENCY IN CHILDHOOD—RICKETS

Rickets is a nutritional disturbance which occurs in children and is due to a deficiency of vitamin D in the diet and lack of sunlight The disease is characterized principally by changes in the calcium content and epiphyseal structures of the bones During the active stages the mineralization of the bones is greatly reduced There is poor absorption of calcium from the intestines with transient lowering of the level of calcium in the blood stream In order to maintain the equilibrium of the blood, calcium is withdrawn from the bones the storehouse of calcium As the

amount of lime in the osseous structures decreases the bones become weakened In infancy the disease is characterized by delay in the appearance of the centers of ossification and flaring of the ends of the shafts of the long bones As the child grows the bones continue to increase in length There is failure of calcification of the greater portion of the new bone resulting in a widening of the space between the diaphyses and the epiphyses The trabeculae are coarse due to the fact that there is much uncalcified osteoid tissue in the bones Pathologic fractures are common Healing of the fractures is slow and callus may not be demonstrable until adequate therapy has been instituted The disease is most prevalent in children one to four years of age but may occur in early infancy and also in the prepuberal period the so called late rickets Bow legs, knock



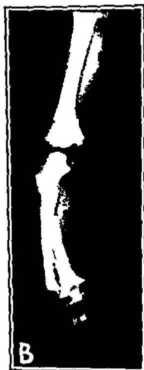


Fig 197 Active Rickets *A* Lower Legs *B* Right Forearm
The metaphyses show cupping fraying and irregularity of outline

knees scoliosis coxa vara and other deformities commonly result In severe clinical rickets death frequently ensues In many cases the child remains dwarfed or crippled for life Vitamin D deficiency varies with the seasons and variability of food containing provitamin D It is important to note that large segments of the population which were formerly afflicted by this disease are now free or almost entirely free because of the advantages of education and better food supply

Roentgen Manifestations The roentgen manifestations are classified in three groups (1) changes in the general architecture of the bone, (2) alterations of the epiphyseal structures and (3) deformities There is osteoporosis with marked thinning of the cortex of the bones The trabeculations are coarser broader and more widely spaced than normally particularly in the metaphyseal regions The epiphyseal ends of the diaphyses show roughening and irregularity and become concave and

widened This has been termed cupping The space between the diaphysis and the epiphysis is increased in width The joint spaces are widened and hazy The epiphyses are small poorly defined and less dense than normal Greenstick and subperiosteal fractures and Looser s lines occur The bones of the skull are extremely thin and cast very faint shadows There may be isolated areas or large segments where the bone appears entirely absent Portions of the head may show bulging due to marked softening The fontanels are large and the sutures wide with indefinite, tapering borders There may be a mosaic pattern with irregular, thin lines extending along the calvaria forming multiple wormian bones Fractures are common

An important characteristic is beading in the epiphyseal regions This is most marked at the wrist and ankle The shafts of most of the long bones show flaring at the junction with the epiphyseal cartilage The addition of new bone at the metaphysis is accompanied by resorption of the flaring ends of the shaft This permits the central more or less cylindrical portion of the shaft to maintain its shape during the period of rapid longitudinal growth There is an addition of new but uncalcified bone at the metaphysis The modeling resorption ceases This is manifested by a club like thickening of the long bones in the metaphyseal regions, the rachitic metaphysis The deformities are most marked in the metaphyseal region where the shaft normally undergoes widening In advanced cases, the bones bend or break under mechanical stress such as body weight a muscle pull Bow legs and knock knees are caused partially by bending of the femur tibia and fibula and in part by a change in the position of the ends of these bones which are joined to the shafts by the markedly thickened epiphyseal cartilage The pelvis is compressed, particularly in its anteroposterior diameter by the weight of the trunk The deformity of the pelvis is a common cause of difficulty in childbirth because the rachitic deformities are never entirely repaired in later life Deformities of the vertebral column such as lordosis kyphosis and scoliosis of varying degree are also common The ends of the ribs, where they join the costal cartilages become enlarged giving rise to the so called "rickety rosary," which in mild cases is only found on the internal surface of the thorax Lateral to these enlargements the softened ribs sink in so as to present a groove passing downward and lateralward on either side of the sternum and the anteroposterior diameter of the chest is increased The ribs affected are the second to the eighth the lower ones being prevented from falling in by the presence of the liver stomach and spleen When the abdomen is distended as it often is in rickets the lower ribs may be pushed outward causing a transverse groove (Harrison s sulcus) just above the costal arch This deformity or forward projection of the

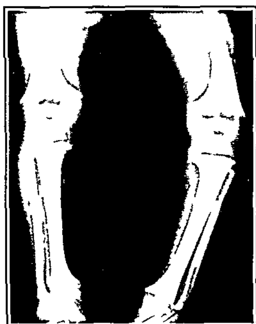
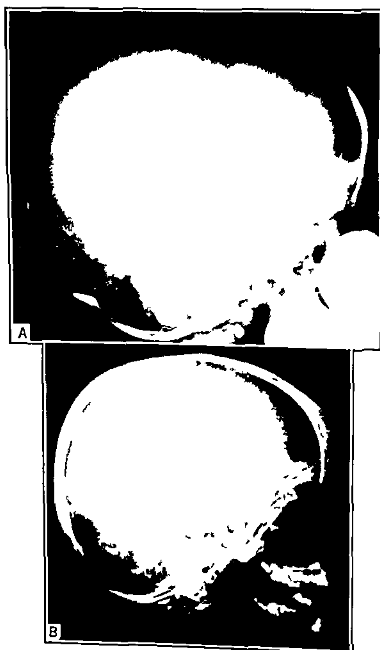


FIG 198 Healing Rickets

sternum, often asymmetrical, is known as pigeon breast and may be taken as evidence of active or old rickets except in cases of primary spinal curvature In some rickety children or adults and also in others who give no history or further evidence of having had rickets an opposite condition obtains The lower part of the sternum and often the xiphoid process as well are deeply depressed backward producing an oval hollow in the lower sternal and upper epigastric regions This is known as funnel breast, it never appears to produce the least disturbance of any of the vital functions

In the later stages, there are hyperostoses, especially in the frontal and parietal regions Grooves form at the coronal and sagittal sutures



F1G 199 The Skull in Rickets A Active stage The sutures are wide and the fontanels large The bones are thin and cast very faint shadows In the sagittal projection numerous Wormian bones were clearly demonstrable B Healed stage There is marked thickening of the frontal and parietal bones with poor definition of the outer tables and diploic spaces

The inner table appears normal. The outer table, the diploic spaces, and the skull margins are obliterated. The vertebrae are narrowed and the disc outlines may become biconcave. The acetabulums are increased in width and coxa vara develops. Bow legs and knock knees are common. There may also be degenerative changes about the knees and the lumbosacral spine. There is increase in the width and density of the long bones. A thin white line extends horizontally across the lower ends of the shafts of the long bones. Periosteal proliferation and elevation may occur. There is bowing with increase in width and density along the concave aspect of the cortex of the long bones. This is in striking contrast to syphilis, the characteristic saber shin being characterized by thickening of the anterior aspect of the corticalis.

Pathogenesis. In order to understand the changes which occur in rickets it is essential to differentiate between the effects of vitamin D deficiency on bone as a tissue and its effect on the bones. The first phase of the development of bone tissue, the formation of the organic matrix by the osteoblasts, is not affected, but the second phase, the mineralization of the matrix or actual calcification, does not ensue. In consequence, an osteoid type of tissue rather than bone is produced. The osteoid tissue is highly resistant to osteoclastic resorption and persists in excessive amounts. The addition of vitamin D to the diet results in rapid transformation of the osteoid into calcified bone. There is failure of calcification not only of the bony matrix but also of the matrix of the hyaline cartilage. Calcification of the cartilage does not terminate abruptly. The proliferation of the cartilage continues far beyond the period when its resorption and replacement by bone should normally have ceased and the thickness of the cartilaginous plate becomes greatly increased. The increase in the thickness of the epiphyseal plates is visible on the roentgenogram and forms one of the important manifestations of the disease. In consequence of the changes in the cartilage there is retardation or temporary cessation of growth dependent on endochondral ossification. Since the long bones are markedly affected, the extremities become short. The length of the trunk is less affected because of the great number of growth centers in the vertebral column. The base of the skull lags in its anteroposterior expansion with apparent bulging of the forehead. As the growth of the mandible is more retarded than that of the facial skeleton, rickets is one of the important factors in the production of facial disharmony. The vertical eruption of the teeth is dependent on the normal growth of the ramus of the mandible in height as this provides the space necessary between the upper and the lower jaws. Impairment of cartilaginous growth at the mandibular condyle results in retardation of the eruption of the teeth. The timing of the tooth eruption is disturbed and accounts for the frequent malposition of the teeth.

The changes in the skull in many instances comprise the earliest manifestations of rickets and are particularly marked because the brain and the skull show a greater rate of growth than other parts of the body at the time that the disease usually has its onset. The growth of the skull occurs in part by endochondral growth at the base but largely by sutural apposition. Resorption proceeds only on the inner surface during the period in which the premorbid calcified bone has not been removed entirely or is not covered by a layer of osteoid tissue. If resorption cannot occur, expansion of the capsule of the growing brain is possible only by sutural growth. The fact that sutural growth must compensate for the

lack of inner resorption is responsible for the increase in width and persistence of the sutures and the fontanels Resorption on the inner surface of the bones of the skull serves two purposes, to enlarge the brain cavity and to flatten the curvatures of the bones which form the cranial vault Failure of resorption and the shift to the sutures of all the changes which contribute to the increase of the cranial capacity lead to persistence of the exaggerated convexity of the central portions of the growing bone There result a general thickening of the bones of the cranial vault and prominence of the frontal and parietal regions with bossing The circumference of the skull of the rachitic infant is increased by the prominence of the bosses and the head appears large The disproportion between the head and body is accentuated by the underdevelopment of the body In many instances the bosses persist throughout life Craniotabes is due to the fact that the bones are extremely thin Flattening develops at points where pressure is greatest and in an infant which lies flat on its back this condition is mechanical in origin

Differential Diagnosis Several diseases must be considered in differential diagnosis In chondrodystrophy the absence of true rachitic changes at the junctions of the epiphyses and the metaphyses and the normal blood chemistry establish the diagnosis In osteogenesis imperfecta there is no difficulty during the period of rapid growth of the tubular bones After closure of the epiphyses, the diseases may simulate each other as in osteogenesis imperfecta there may be generalized osseous rarefaction flaring of the metaphyses and extensive deformities due to softening of the bone and multiple fractures The presence of blue scleræ and the normal blood serum phosphorus levels usually establish the diagnosis Hyperparathyroidism in the primary form is extremely rare in infants and children During adolescence, the pseudorachitic changes in the metaphyses can be differentiated from true rickets on the basis of the blood studies and the presence of areas of cystic rarefaction in the long bones In renal rickets the bone changes due to chronic renal insufficiency are actually due to secondary hyperparathyroidism Both diseases occur in infancy and childhood, are associated with dwarfism and may produce identical roentgen manifestations The diagnosis depends on renal function tests and studies of the blood chemistry In renal rickets the serum phosphorus level is high whereas in refractory rickets it is low The rare form of late rickets termed the Fanconi syndrome is believed to be due to degenerative changes in the renal tubular epithelium Both diseases are characterized by an hereditary tendency and hypophosphatemia In the Fanconi syndrome there is renal glycosuria and aminoaciduria neither of which is found in rickets Other forms of rickets such as late rickets and rachitic changes occurring in certain forms of pancreatic, hepatic or intestinal insufficiency and congenital biliary atresia may be very difficult to differentiate Syphilis may be confused with the late stages of rickets Late syphilis is characterized by the presence of the sabre shin with bowing and thickening along the anterior aspect of the cortex while in rickets the cortical thickening involves the concavity of the bone curvature

Double Contour Cupping and Spurring in the Long Bones of Infants There not infrequently occurs in the long bones of infants a double contour effect with duplication of the cortical shadow The condition is characterized by the presence of a continuous linear shadow adjacent to the diaphysis along its external lateral aspect and separated from it by

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The changes in the skull in many instances comprise the earliest manifestations of rickets and are particularly marked because the brain and the skull show a greater rate of growth than other parts of the body at the time that the disease usually has its onset The growth of the skull occurs in part by endochondral growth at the base but largely by sutural apposition Resorption proceeds only on the inner surface during the period in which the premorbid calcified bone has not been removed entirely or is not covered by a layer of osteoid tissue If resorption cannot occur, expansion of the capsule of the growing brain is possible only by sutural growth The fact that sutural growth must compensate for the

lack of inner resorption is responsible for the increase in width and persistence of the sutures and the fontanels Resorption on the inner surface of the bones of the skull serves two purposes to enlarge the brain cavity and to flatten the curvatures of the bones which form the cranial vault Failure of resorption and the shift to the sutures of all the changes which contribute to the increase of the cranial capacity lead to persistence of the exaggerated convexity of the central portions of the growing bone There result a general thickening of the bones of the cranial vault and prominence of the frontal and parietal regions with bossing The circumference of the skull of the rachitic infant is increased by the prominence of the bosses and the head appears large The disproportion between the head and body is accentuated by the underdevelopment of the body In many instances the bosses persist throughout life Craniotabes is due to the fact that the bones are extremely thin Flattening develops at points where pressure is greatest and in an infant which lies flat on its back this condition is mechanical in origin

Differential Diagnosis Several diseases must be considered in differential diagnosis In chondrodystrophy the absence of true rachitic changes at the junctions of the epiphyses and the metaphyses and the normal blood chemistry establish the diagnosis In osteogenesis imperfecta there is no difficulty during the period of rapid growth of the tubular bones After closure of the epiphyses, the diseases may simulate each other as in osteogenesis imperfecta there may be generalized osseous rarefaction, flaring of the metaphyses and extensive deformities due to softening of the bone and multiple fractures The presence of blue sclerae and the normal blood serum phosphorus levels usually establish the diagnosis Hyperparathyroidism in the primary form is extremely rare in infants and children During adolescence the pseudorachitic changes in the metaphyses can be differentiated from true rickets on the basis of the blood studies and the presence of areas of cystic rarefaction in the long bones In renal rickets the bone changes due to chronic renal insufficiency are actually due to secondary hyperparathyroidism Both diseases occur in infancy and childhood are associated with dwarfism and may produce identical roentgen manifestations The diagnosis depends on renal function tests and studies of the blood chemistry In renal rickets the serum phosphorus level is high whereas in refractory rickets it is low The rare form of late rickets termed the Fanconi syndrome is believed to be due to degenerative changes in the renal tubular epithelium Both diseases are characterized by an hereditary tendency and hypophosphatemia In the Fanconi syndrome there is renal glycosuria and aminoaciduria neither of which is found in rickets Other forms of rickets such as late rickets and rachitic changes occurring in certain forms of pancreatic, hepatic or intestinal insufficiency and congenital biliary atresia may be very difficult to differentiate Syphilis may be confused with the late stages of rickets Late syphilis is characterized by the presence of the sabre shin with bowing and thickening along the anterior aspect of the cortex, while in rickets the cortical thickening involves the concavity of the bone curvature

Double Contour, Cupping and Spurring in the Long Bones of Infants There not infrequently occurs in the long bones of infants a double contour effect with duplication of the cortical shadow The condition is characterized by the presence of a continuous linear shadow adjacent to the diaphysis along its external lateral aspect and separated from it by

an area of increased radiance measuring about 1 mm in width It is not laminated and is limited to the diaphyseal portion of the bone The double contour appearance is most frequent among premature infants of low birth weight The more marked the change initially, the longer it persists In a group of apparently healthy infants studied by Hancox *et al*, 23 per cent showed the condition Once the condition disappeared, it did not reappear It was not noted to develop later in instances in which the initial roentgen studies had been normal



Fig 200 Double Contour and Cupping The lower ends of the diaphyses of the radius and ulna show cupping There is a continuous line at the margin of the diaphysis of each of the long bones imparting a double contour appearance to the long bones

Glaser performed roentgen studies of 100 prematurely born babies Roentgenograms of the long bones, wrists, and ankles were taken at monthly intervals until the eighth month of life, thus obtaining a series of 800 roentgenograms The surfaces of the long bones of the forearm and leg were frequently outlined by a double contour which simulated periosteal thickening or elevation and cortical hypertrophy The lines were present in the distal parts of all of the bones of the extremities or on only one or more They were found most frequently in the ulna and with decreasing frequency in the radius, tibia and fibula In many instances they first became visible in one bone and after an interval of two or three months were noted in other bones The changes affected the entire length of the metaphysis or were limited to one portion of the bone, extending parallel to the periosteum for a variable distance and fading into the subcortical structures In many of the infants there was a peculiar appearance of the epiphyseal line with certain alterations which might easily be interpreted as signs of disease Normally the epiphyseal plate assumes the form of a straight line If the roentgen rays traverse the epiphyseal plate at an angle other than a right angle it may appear concave or convex and the edges become elongated producing an appearance which resembles the spurring lipping or cupping of scurvy or rickets The cupping and spurring were first noted during the third or fourth month of life and diminished at about the seventh to eighth month Originally the alterations in the bones were interpreted as being due to rickets scurvy or congenital syphilis It was later decided that the changes were most probably physiologic in origin Glaser made comparative studies in children not premature and found somewhat similar appearances The changes were less marked and of less frequent occurrence in the full term children Studies of the blood calcium, phos

phorus and phosphatase and serologic studies revealed no abnormalities Double contour cupping and spurring may occur in normal children These changes should be considered a normal roentgen appearance of the long bones during the period of growth and should not be misinterpreted as signs of a pathological process They have been noted in healthy premature and full term babies Histopathologic examination of sections of the bones of infants presenting these manifestations during life and who died of unrelated conditions indicate that they are normal and disappear without treatment or change in diet There is no correlation with lues rickets or other disease

The Occurrence of Rickets in Association with Other Skeletal Disease in Infants and Children

The Simultaneous Appearance of Rickets and Scurvy Involvement of the skeleton by rickets in association with other disease processes of the skeleton is relatively common Rickets may occur in combination with congenital syphilis scurvy lead poisoning erythroblastic anemia and osteogenesis imperfecta The diagnosis of rickets in association with other diseases may be difficult Scurvy and rickets are much less common than twenty or thirty years ago The etiology of these diseases is now definitely understood and when they do occur it is because of ignorance or lack of adequate preventive and curative treatment In the past it was relatively common to find rickets and scurvy simultaneously In many instances the roentgen study does not demonstrate the rickets unless the disease is well established and the diagnosis must be made on the basis of a history of vitamin deficiency the season of the year when the patient comes under observation and the results of clinical and roentgen examination aided by measurements of the calcium inorganic phosphorus, and phosphatase of the serum This is due to the fact that some manifestations occur in both diseases with rarefaction and thinning of the cortex, spreading or cupping of the diaphyseal ends of the long bones lateral spurs rosary at the ends of the ribs, and a zone of preparatory calcification Despite the fact that the signs of scurvy tend to obscure those of rickets unless the latter disease is well developed and severe the diagnosis can be established by careful study of the roentgenograms in most instances The rarefaction of the cortex of the affected bones is the most significant sign in roentgen diagnosis A moth eaten type of rarefaction is more characteristic of rickets In many instances this predominates An important aid in diagnosis is the presence of the zones of preparatory calcification In scurvy this zone is denser and at times broader than normal In rickets particularly in the early stage and prior to the development of healing this area is hazy frayed out and poorly defined When the two diseases occur simultaneously the osteoid zone of rickets can be identified by the presence of a marked increase in the width of the clear space between the epiphyseal center and the end of the diaphysis The ring shadow about the epiphyseal center remains intact and indicates the presence of scurvy although at the end of the diaphysis the zone of preparatory calcification presents a frayed out appearance Spreading or widening of the diaphyseal ends of the bones cannot be considered a definite manifestation of either of the diseases as it may occur in both In scurvy the changes in the periosteum may be of two types In one form of the disease there is a narrow, triangular

shadow with its base at the metaphysis and extending for a variable distance along the shaft Another variety is characterized by periosteal shadows which are associated with more severe hemorrhages

The Simultaneous Appearance of Rickets and Congenital Syphilis When these two diseases occur simultaneously, it is usually in early infancy In these instances the diagnosis of rickets is established by slight fraying of the zones of preparatory calcification slight widening of the diaphyseal ends, slight thickening of the periosteum in the region of the borders of the zone particularly in the radius and ulna where the interosseous muscles are inserted and slight decalcification of the shaft The periosteal changes in the two conditions are practically identical Rickets developing in a case of congenital syphilis may completely obscure the signs of osteochondritis which are so common in lues Antiluetic therapy may cause an apparent change in the underlying or associated rickets

Rickets and Lead Poisoning Chronic lead poisoning may be superimposed on or occur in association with rickets In some instances the lead zones at the diaphyseal ends of the long bones may not be demonstrable The presence of marked stippling of the red cells is an important aid in diagnosis The presence of a wide osteoid zone between the diaphyseal ends and the centers of ossification together with the zones of preparatory calcification in the long bones indicative of rickets may obscure the changes caused by the lead In lead poisoning the lead line may fail to develop in the presence of associated rickets due to the fact that calcification of the preparatory cartilage has ceased In lead poisoning there is excessive calcification of the preparatory cartilage while in rickets the deposition of calcium and lead at the same sites cannot take place In severe rickets the preparatory cartilage does not absorb calcium or lead and the characteristic lead line therefore will be absent There results a layer of radiolucent uncalcified cartilage and osteoid in the terminal segment of the bone rather than the densely calcified cartilage which develops in plumbism without associated rickets and is responsible for the lead line in the roentgenogram This may be due to the fact that the lead ingested has produced a low phosphate level When the rickets heals the lead line may appear in the bone

Rickets and Scurvy Occurring in a Case of Osteogenesis Imperfecta Bromer reports a case of a Negro male infant three months of age in whom roentgenograms showed evidence of pathological fractures, atrophy of the long bones and a parchment like skull There was no blue tinge to the sclera In association with these changes the bones of the forearm showed decalcification of the shafts of the radius and ulna bilaterally slight flaring of the diaphyseal end of the right radius and an osteoid zone distal to the frayed out zone of preparatory decalcification The signs of rickets disappeared with the feeding of vitamin D Six months later the patient presented the characteristic signs of scurvy There were subperiosteal hemorrhages ring shadows about the epiphyseal centers, lateral spurs and slightly increased density of the zones of preparatory calcification The roentgen signs of scurvy disappeared after the administration of vitamin C

Vitamin D Resistant Rickets Refractory Rickets

Rickets resistant to therapy with vitamin D the so called refractory rickets, is a rare metabolic disorder of childhood which differs from ordi

nary infantile rickets in that large doses of vitamin D fail to bring about cure or prevent recurrences Since the use of concentrated preparations of vitamin D has reduced the incidence of florid infantile rickets to practically the vanishing point in the United States the refractory variety has assumed a position of increasing importance It is more common than previously realized and many cases of the disease are unrecognized at present The blood chemistry in patients with refractory rickets is practically identical with that in infantile rickets and is characterized by normal serum calcium low serum phosphorus and elevated serum phosphatase An important difference lies in the fact that the serum phosphorus level remains persistently low despite intensive therapy Also decreased urinary calcium excretion is not as prominent a feature as in ordinary rickets There is increased fecal excretion of calcium and phosphate which has been shown to be due to decreased absorption of these substances from the intestinal tract The reason for this aberration of intestinal absorption in the presence of what is normally more than a sufficient amount of vitamin D is the unknown factor in refractory rickets The condition is very apt to occur in several members of the same family, which indicates that the abnormality may be inherited It is important in that it represents one of the most common causes of dwarfism

In the typical case of unrecognized refractory rickets the onset occurs early in the post infantile period and the condition persists to late childhood A roentgen diagnosis of rickets is usually made but is not considered correct when it is learned that the patient has received an adequate supply of vitamin D This is further intensified when the usual forms of treatment do not result in a cure Operative procedures principally osteotomies are of no avail because the rickets continues active and the deformities recur In some instances the disease is arrested spontaneously at puberty but in others it persists into adult life as osteomalacia and is termed adult rickets There is no evidence of kidney liver or gastro intestinal disease It appears that the vitamin D which the patient receives is absorbed There is no response to treatment until a threshold level has been reached and above this level the response is normal Manifestations of toxicity appear if the vitamin D intake becomes too excessive An adequate response by the patient is indicated by increased urinary calcium and decreased fecal calcium and in the blood by increased inorganic phosphorus normal or elevated calcium and a gradual decline in alkaline phosphatase In association with this chemical response there are roentgen evidences of healing with growth of the patient at the rate of about 1 *cm* a month Once healing has been established the maintenance dose of vitamin D may be greatly decreased It is now well established that vitamin resistant rickets is a distinct clinical entity

Vitamin resistant rickets is differentiated from renal acidosis and the Fanconi syndrome by decreased urinary calcium excretion, the absence of glucose, albumin and amino acids in the urine and the presence of normal plasma electrolytes The disease is related to renal acidosis due to tubular disease and the Fanconi syndrome However parathyroid hyperplasia is known to exist in rickets and the increased phosphorus excretion in rickets is probably due to parathyroid activity Since vitamin therapy increases the serum calcium level, there should be decreased parathyroid activity and decreased phosphorus excretion There is a strong familial tendency to the disease In the past it was classified

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Albright and his co workers classify osteomalacia into four types (1) osteomalacia with normal phosphatase, (2) chemical osteomalacia with high phosphatase, (3) Milkman s syndrome and (4) advanced osteomalacia They suggest the following classification of the causes of osteomalacia

A *Insufficient Vitamin D* (1) Dietary lack This is rare in the United States as the average diet contains adequate amounts of vitamin D and the food intake is amply supplemented by the effects of sunshine The condition is not infrequently seen in India and China due to voluntary imposition of an abnormal diet During and after World War I dietary restrictions in certain instances resulted in the development of many cases of osteomalacia in the Central European countries This



Fig 201 Fig 202

Fig 201 Osteomalacia There is very marked generalized decalcification of the bones The cortex of the long bones is decreased in thickness and density The first impression on observing the roentgenogram is that the film is underexposed but this is due to the marked decalcification of the bones

Fig 202 Osteomalacia The bones of the pelvis show advanced osteoporosis The pelvic cavity is markedly narrowed

hunger osteopathy due to lack of vitamin D responds to the administration of cod liver oil (2) Gastrointestinal disorders Vitamin D is fat soluble and is not absorbed in certain diseases, particularly sprue and steatorrhea The associated diarrhea with rapid passage and evacuation of intestinal contents adds to the difficulty of absorption of calcium and osteomalacia not infrequently develops in such cases Addiction to laxatives especially oils and mineral salts *which have a tendency* to form insoluble calcium salts in the intestinal tract and chronic achlorhydria with reduction of absorption of calcium may also act as causative factors The most common cause of hypovitaminosis D in adults in the United States is steatorrhea There is a loss of the fat soluble vitamins A D, K and E in the stools and a deficiency of calcium absorption from the intestinal tract In consequence there is improper calcification of bone

B *Osteonephropathy* In this group are included renal rickets DeToni Fanconi s syndrome, and idiopathic hypercalcariuria These entities are discussed on pages 401 410 and 413

incorrectly as achondroplasia, dyschondroplasia chondrodysplasia, or a combination of these with rickets The diagnosis of vitamin resistant rickets is established definitely by studies of the urine and blood chemistry

During the active stage of the disease, the patients present the usual sign of active rickets with enlargement of all the palpable epiphyses, Harrison's groove, the rachitic rosary, and bossing in the frontal region The patients are very small in stature due to lack of growth, this varies with the severity of the disease and is most marked in the lower extremities There are severe deformities in the lower legs with bow legs and knock knees There may be a double deformity with a bow on one side and a knock knee on the other Marked anterior bowing of the femurs and tibia may ensue These tend to recur after correction by osteotomy There is generalized rarefaction and coarsening of the trabecular pattern of various bones with irregular fraying of the epiphyseal plates widening of the spaces between the epiphyseal ossification centers and the shafts and cupping of the ends of the shafts There are also green stick fractures pseudofractures and angular deformities of the extremities There is apt to develop rickets in unusual locations such as the angles of the scapulas the elbows and the ischiopubic synchondroses The humeri may appear short and broad The thoracic cage is usually deformed The lamina dura of the teeth is absent or poorly defined

VITAMIN D DEFICIENCY IN ADULTS

Osteomalacia

Osteomalacia is a term applied to a disorder of adults characterized by the presence of abnormal amounts of osteoid tissue in the bones The diagnosis can be established with definiteness only by histopathologic study of the bone Clinical studies roentgen examination and the response to therapy afford very important evidence as to the character and extent of the disease It is very rare in the United States and is apt to be unrecognized The pathology is similar to that of rickets The disease occurs after active bone growth has ceased and the manifestations are those resulting from replacement of the bone trabecula by fibrous or osteoid tissue It is a condition which occurs most frequently in lactating or pregnant women at times of greatly increased calcium demand but is not limited to females There is excessive calcium excretion and the blood calcium and phosphorus levels are disturbed Bone which was previously normal is deprived of its calcium and is replaced by osteoid tissue The calcium lost by the bones is excreted in the feces and the bones fail to recalcify The effects of vitamin D deficiency during the period of skeletal growth are markedly different than after growth has ceased In the latter period the formation of new bone does not contribute to a significant degree to the growth of the skeleton but is an important factor in the process of replacement of overaged bone by young vital bone In consequence vitamin D deficiency in the growing individual produces rickets while in the adult it results in osteomalacia An important additional factor is the increased demand on the body economy during pregnancy and lactation Any condition which causes severe depletion of the calcium stores of the body may be a cause of osteomalacia The depletion may develop because of insufficient absorption or excessive loss of calcium



Fig 203 Milkman's Disease *A* The right femur There is very marked decalcification of the entire shaft of the femur with extreme thinning of the cortex Thin linear fractures with partial filling in of the fracture lines are faintly visualized (arrow) *B* The left forearm The extreme osteoporosis is well shown There are transverse fractures of the radius and ulna with no separation or displacement of the fragments

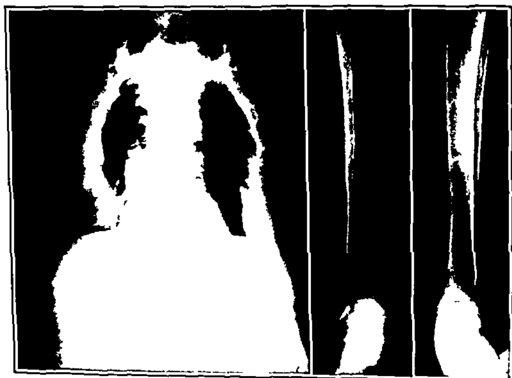


Fig 204 Milkman's Disease There is marked osteoporosis of the bony thorax with narrowing of the interspaces and inward displacement of the ribs

C *Transient Osteomalacia Subsequent to Removal of a Parathyroid Adenoma* After removal of a parathyroid adenoma, there is a sudden drop in serum calcium with disturbance of the mineral balance to a sufficient degree to interrupt the processes of calcification This is a temporary, self limited situation which is not of clinical significance and does not constitute a true form of osteomalacia

Roentgen Manifestations On roentgen study, there is markedly increased radiance of the bones with marked cortical thinning and widening of the medullary cavity The margins of the bone are poorly defined and may be barely distinguishable from the adjacent soft tissues The trabeculae are very indistinct The pelvis is narrow and contracted There is a severe lordosis The long bones are bowed and fracture easily Callus formation is very poor There may be widening of the diaphyses and cystic cavities may develop within the bones If healing takes place, deformities persist in the recalcified bones There may be great difficulty in differentiating the condition from osteitis fibrosa cystica

Milkman's Syndrome

In 1930 Milkman reported a case of pseudo fractures in a forty-year old white female with multiple symmetrical incomplete fractures which had gradually been increasing in number over a period of five years Looser had shown previously that hunger osteopathy, late rickets and osteomalacia are similar in nature and differ in their clinical manifestations only because of the different age periods in which they occur Milkman classed his case with those of Looser and stressed that the basic etiological factor was unknown It was his opinion that the condition was not osteomalacia and that he was dealing with a new clinical entity He felt that the zones of radiolucency manifested as pseudofractures roentgenographically were due to a trophic disturbance associated with a particular pattern of bone absorption which is not typical of any known disease entity

Milkman's syndrome is considered a form of osteomalacia due to a disorder of calcium and phosphorus metabolism The bony matrix is formed normally to a certain point but the calcium salts are not deposited in the newly formed osteoid tissue Osteomalacia is distinguished from osteoporosis by the fact that in the latter the osteoblasts lay down too little bony matrix although the matrix which is formed is calcified normally Rickets presents all the characteristics of osteomalacia and in addition changes at the growing epiphyseal cartilage particularly calcification of the zone of provisional calcification For this reason osteomalacia has been termed adult rickets

The principal clinical manifestations of Milkman's syndrome are *intermittent pain in the back and legs easy fatiguability a waddling gait* and difficulty in changing position The onset is insidious The patients may appear to be in good nutritional condition and of excellent constitution despite the presence of widespread and advanced change in the bones However in the late stages they become bedfast and the body length decreases because of the skeletal deformities The disease occurs primarily in females all of the recorded cases with one exception having been in women Laboratory studies reveal normal findings Multiple fractures are common, in one instance more than forty fractures being described Autopsy studies have disclosed no pathognomonic findings in the disease

The important manifestations comprise osteoporosis or osteosclerosis and metastatic calcifications The abnormal depositions of calcium have been reported in the major arteries, the vessels of the extremities, the heart, the lungs, the alveoli and bronchi, the kidneys, the gastric mucosa, the pancreas, the articular bursas, the skin, the subcutaneous tissues, the muscles the sclera the falx, the choroid and the dura In many instances the calcific depositions occur as manifestations of other disease syndromes or as incidental findings in otherwise normal individuals It is, therefore, important that the diagnosis be made on the basis of the history and the clinical findings Osteoporosis similar to that in osteitis fibrosa may occur In other cases the changes in the bones resemble osteopetrosis The skull is dense and thick, especially in the basal and frontal bones and the

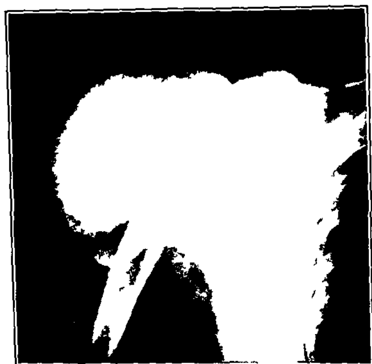


Fig 205 Hypervitaminosis D There are multiple amorphous areas of calcific density in the soft tissues about the shoulder There is generalized osteoporosis The patient had been taking large amounts of Vitamin D for the treatment of arthritis

orbital roof The ribs and vertebrae are opaque The flat bones of the pelvis are dense and wide The long bones are markedly osteosclerotic The process involves the diaphysis with cortical expansion The medullary canal is narrowed but not obliterated The epiphyseal nuclei and the small bones of the hands and feet may show a radiant peripheral zone surrounding a sclerotic center

Pathogenesis The effects of overdosage of vitamin D are not clearly understood Some observers are of the opinion that osteoporosis is an early stage of vitamin D intoxication while the later stages are characterized by osteosclerosis The explanation of these contradictory findings according to Weinman and Sicher is based on the effect of excessive amounts of vitamin D on the kidney If the kidney is not affected or is injured only slightly, the overdosage of vitamin D results in the development of osteosclerosis Damage to the kidney causes renal hyperpara

The prognosis is dependent on the response to therapy of the underlying causative factors. No etiologic agent has been established. The occurrence of the condition in pregnancy is rare. It has been reported as occurring in puerperal osteomalacia.

Roentgen Manifestations. On roentgen examination the disease is characterized primarily by generalized osteoporosis and a multiplicity of symmetrical fractures which show inadequate healing. In the early phases there are focal areas of osteoporosis which progress to linear areas of rarefaction. These areas have been termed Looser's zones and are generally referred to as pseudofractures. Above and below the lesions the bone appears normal except for osteoporosis. With progression of the disease there is displacement at the sites of the fractures with resultant deformities, particularly coxa vara. Displacement is rare and the lesions appear to remain static by roentgen examination with little or no evidence of healing. The periosteum is not affected and the outline of the bone is unchanged. The Looser's zones measure 1.0 mm. to 1.0 cm. in width, extend across the bone, and present a clear band as though the bone had been removed completely. Signs of regression are rare and are manifested by two dense bands, one on each side of the clear zone.

Differential Diagnosis. Other diseases in which Looser's zones or pseudofractures have been described and which must be considered in differential diagnosis comprise osteomalacia, rickets, late rickets, renal rickets, celiac disease, chronic idiopathic steatorrhea, intestinal infantilism, sprue, non-tropical sprue, osteogenesis imperfecta, fragilitas ossium, hyperthyroidism, hyperparathyroidism, Paget's disease, bone dystrophy secondary to adrenopituitary dysfunction, severe chronic acidosis as in hyperglycemia, congenital syphilis, osteomyelitis, osteopetrosis, march fracture, cough fracture, and the blood dyscrasias.

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Milkman, L. A.: Pseudofractures (Osteopathy, Late Rickets, Osteomalacia). Am. J. Roent. *24* 29 1930. Also Multiple Spontaneous Idiopathic Symmetrical Fractures. Am. J. Roent. *32* 622 1934.

VITAMIN D INTOXICATION

The use of large doses of vitamin D in the treatment of arthritis and other disease is a dangerous practice which may result in permanent damage and death. Renal function and calcium metabolism studies are essential in all patients receiving massive vitamin D therapy, any deviation constituting an absolute contraindication to the continuation of the treatment. Massive vitamin D administration need not be prolonged to cause toxicity as patients have become severely incapacitated after receiving 150,000 units of vitamin D daily for only seven weeks. Cessation of vitamin D administration in the presence of toxic symptoms usually results in clinical improvement with partial or complete disappearance of the toxic effects. In all instances of hypercalcemia or unexplained renal disease, vitamin D intoxication should be considered in the differential diagnosis. Toxic effects from vitamin D occur only after very high dosage. Damage to the kidneys is common. Death has resulted in many instances.

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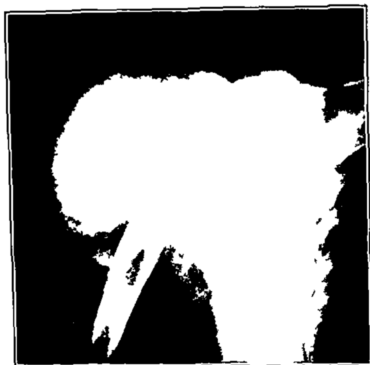


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The prognosis is dependent on the response to therapy of the underlying causative factors No etiologic agent has been established The occurrence of the condition in pregnancy is rare It has been reported as occurring in puerperal osteomalacia

Roentgen Manifestations On roentgen examination the disease is characterized primarily by generalized osteoporosis and a multiplicity of symmetrical fractures which show inadequate healing In the early phases, there are focal areas of osteoporosis which progress to linear areas of rarefaction These areas have been termed Looser s zones and are generally referred to as pseudofractures Above and below the lesions the bone appears normal except for osteoporosis With progression of the disease there is displacement at the sites of the fractures with resultant deformities, particularly coxa vara Displacement is rare and the lesions appear to remain static by roentgen examination with little or no evidence of healing The periosteum is not affected and the outline of the bone is unchanged The Looser s zones measure 1 0 mm to 1 0 cm in width extend across the bone and present a clear band as though the bone had been removed completely Signs of regression are rare and are manifested by two dense bands, one on each side of the clear zone

Differential Diagnosis Other diseases in which Looser s zones or pseudofractures have been described and which must be considered in differential diagnosis comprise osteomalacia rickets late rickets renal rickets, celiac disease, chronic idiopathic steatorrhea, intestinal infantilism, sprue, non tropical sprue osteogenesis imperfecta fragilitas ossium hyperthyroidism hyperparathyroidism Paget s disease bone dystrophy secondary to adrenopituitary dysfunction severe chronic acidosis as in hyperglycemia congenital syphilis osteomyelitis osteopetrosis march fracture, cough fracture and the blood dyscrasias

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VITAMIN D INTOXICATION

The use of large doses of vitamin D in the treatment of arthritis and other disease is a dangerous practice which may result in permanent damage and death Renal function and calcium metabolism studies are essential in all patients receiving massive vitamin D therapy any deviation constituting an absolute contraindication to the continuation of the treatment Massive vitamin D administration need not be prolonged to cause toxicity as patients have become severely incapacitated after receiving 150 000 units of vitamin D daily for only seven weeks Cessation of vitamin D administration in the presence of toxic symptoms usually results in clinical improvement with partial or complete disappearance of the toxic effects In all instances of hypercalcemia or unexplained renal disease, vitamin D intoxication should be considered in the differential diagnosis Toxic effects from vitamin D occur only after very high dosage Damage to the kidneys is common Death has resulted in many instances

OSTEONEPHROPATHY

Osteonephropathy is a general term used to designate a variety of closely related conditions characterized by bone changes which occur in association with disease of the kidneys. There are four clinical syndromes which manifest themselves by alterations of the osseous and endocrine systems (1) Renal rickets, (2) Fanconi syndrome, (3) nephrocalcinosis and (4) idiopathic hypercalcinuria. The most common member of the group is renal rickets.



FIG 206 Renal Rickets There is very marked osteoporosis of the skull with cortical thinning. Multiple areas of increased radiance varying in size from a few millimeters to almost 1 cm in diameter are scattered irregularly throughout the skull. Some of these areas have a honeycomb appearance

Renal Rickets

The association of chronic renal insufficiency with bone changes was first recognized in 1883. The disease is not rare. It is also known as renal dwarfism, renal hyperparathyroidism, renal osteodystrophy, renal osteitis and renal nanism.

Etiology There are various theories as to the cause of renal rickets and the disease has variously been ascribed to a disorder of the renal tubules, the parathyroid glands and the pituitary gland. It is now generally accepted that renal rickets is the result of renal insufficiency and that the other phenomena are of significance only secondarily. Primary hyperparathyroidism results in damage to the kidney due to the deposition of calcium in the renal parenchyma. Unless the adenoma is removed, marked insufficiency of the kidney results. In consequence, there develop changes in the bone and the blood chemistry which are similar to those in renal rickets. Both primary and secondary hyperparathyroidism may be present in the same individual. The parathyroid adenoma may eventually cause insufficiency of the kidney and this results in hyperphosphatemia and hyperplasia of the remaining parathyroids.

thyroidism and a form of osteitis fibrosa cystica generalisata with osteoporosis ensues. Dosages which cause moderate damage to the kidney bring about an exaggeration of the normal effect of vitamin D ingestion. The absorption of calcium and phosphorus in the intestinal tract is increased and the secretion of phosphate by the urine is decreased by the inhibitory effect of the vitamin on the parathyroids. In consequence of the rise of the level of calcium and phosphorus in the blood there is new bone formation and this is followed by the deposition of calcium phosphates in the blood vessels, the ligaments, and the kidneys. The calcification of periosteal and endosteal connective tissue, the presence of calcium deposits in the marrow spaces and the osteosclerosis may be responsible for the apparently increased densities of the bones as visualized roentgenographically. When the tolerance level for vitamin D has been exceeded, damage to the kidney complicates the picture. There is retention of phosphates in the body and the phosphates exceed the amounts of calcium present. In an attempt to eliminate the relative excess of phosphorus, the parathyroid glands undergo hypertrophy. For a period of time the antagonistic action of the inhibitory factor of vitamin D and the stimulating factor of hyperphosphatemia result in a confusing picture. In the later stages the damage to the kidneys and the resultant hyperparathyroidism are the determining factors in the changes which develop in the bones and the symptoms of generalized osteitis fibrosa cystica usually develop.

The Relationship of Parathyroid Hormone and Vitamin D The parathyroid hormone and vitamin D have a common function, *that is,* to maintain the blood calcium level. However, they accomplish this by entirely different mechanisms, the parathyroid hormone and vitamin D being antagonistic. The action of vitamin D is to increase the utilization of the external or dietary resources of calcium, while the parathyroid hormone mobilizes calcium from the internal deposits, that is the bones. Vitamin D maintains or causes an elevation of the blood calcium level by increase in the absorption of calcium from the intestinal tract. By mobilizing the external resources vitamin D restricts the withdrawal from the skeleton of the emergency supplies of calcium and acts to inhibit the action of the parathyroid gland. The parathyroid hormone increases or maintains the blood calcium level by withdrawing calcium from the skeleton through the promotion of osteoclastic resorption. The excessive phosphorus liberated by osteoclasis is eliminated by the kidney. The increase in urinary secretion of phosphorus is brought about by the direct action of the parathyroid hormone on the kidney. The parathyroid hormone exerts an inhibiting influence on vitamin D, particularly its calcifying action. Normally the antagonistic actions of vitamin D and the parathyroid hormone result in an equilibrium.

phate form This leads to a calcium deficient rickets Albright is of the opinion that the bowel changes in renal rickets are not directly connected with the secondary hyperparathyroidism but are dependent on the associated acidosis It is not always possible to correlate the type and severity of the bone disease with the degree of secondary hyperparathyroidism Also there is no direct relationship between the degree of acidosis, the elevation of the blood urea nitrogen, or the extent of retention of phosphate and the type and severity of the bone changes

Pathology The changes in the kidneys are widely variable but are associated with marked insufficiency of the glomeruli and the tubules The abnormalities in the bone cannot be differentiated from those which occur in generalized osteitis fibrosa associated with primary hyperparathyroidism Therefore, the condition may be termed renal osteitis fibrosa generalisata In children, there occur lesions of the epiphyses which closely resemble rickets on roentgen study but are not those of true rickets on microscopic examination The four parathyroid glands are usually involved and the size of the glands may vary widely

Clinical and Laboratory Manifestations The disease occurs in infancy, childhood or adolescence and is manifested by varying degrees of retardation of growth, changes in the bones and chronic renal insufficiency The renal insufficiency results in severe acidosis azotemia and hyposthenuria In many instances the patient dies in uremia On clinical examination the most marked change is failure to grow and gain in weight There may be excessive thirst and polyuria but these are not constantly present There are marked deformities of the bone such as genu valgum Dwarfism is common The patients are short small and underweight Renal rickets shows no special racial incidence It is not seen in Negroes in whom true rickets is common The average age of those affected is about eight to ten years There is retarded growth often of a very marked degree, and walking eruption of the teeth and sexual development are delayed On clinical examination many of the stigmata of true rickets are apparent There are large parietal bosses, widening of the epiphyses, flaring and beading of the costochondral junctions Harrison's grooves deformities of the knees and flat feet The laboratory findings are typical and establish the diagnosis The serum calcium is normal or slightly low the serum phosphorus is high, and the alkaline phosphatase is elevated There is a reversal of the normal calcium phosphorus ratio

Roentgen Manifestations The bone changes in renal rickets are widely variable and are usually atypical They resemble a combination of rickets and hyperparathyroidism, one or the other usually predominating The single roentgen manifestation which is invariably present consists of a generalized demineralization of a coarse trabecular type which involves all of the long bones The changes may be divided into two groups In one type there is a rachitic picture which cannot be differentiated from ordinary infantile rickets In the second form there is a woolly change which is characteristic and establishes the diagnosis This is more common but is not as symmetrical as in ordinary rickets The metaphyses present a stippled woolly moth eaten appearance with irregular ossification The metaphyses are similar to those in ordinary rickets but are less markedly splayed In association with the woolly type of change, there occur subperiosteal erosions in the regions of the metaphyses This is considered one of the most significant features in the roentgen diagnosis of renal rickets Secondary displacement of the epiphyses may occur in

Various types of kidney lesions may be the cause of renal rickets These can be divided into three main groups (1) infection or inflammation such as chronic glomerulonephritis and pyelonephritis, (2) congenital anomalies such as polycystic disease, hypogenesis of the kidney and similar malformations, and (3) obstructions of the renal system caused by valves of the urethra and strictures of the urethra bladder neck and ureter There is no data as to which type of kidney disease is the most common etiologic factor In most instances there is a small atrophic kidney due to long standing chronic infection There is some evidence to indicate that the condition is due to disease originating in the kidneys in infancy and childhood, most commonly congenital anomalies such as bilateral hydronephrosis and polycystic kidneys



Fig 207 Renal Rickets There is marked decalcification of the bones The diaphyseoepiphyseal junctions are markedly abnormal with a wide band of increased radiance irregularity of outline *and mottling in this region The femurs show thinning of the cortex There are areas of radiance* along the lateral margins of the lower portion of the ilium bilaterally extending into the superior aspects of the acetabulums The bones are small and underdeveloped

Pathogenesis As a result of the severe damage to the kidneys, there is inability to excrete phosphorus with retention of inorganic phosphorus There is decrease in the level of the serum calcium as an adjustment to the high serum level of phosphorus In consequence there develops a hyperplasia of the parathyroid The calcium is withdrawn from the bones in an effort to maintain the level of the calcium in the serum However the attempted physiological compensation is unsuccessful because of the insufficiency of the kidneys Normally the hyperplasia of the parathyroid would cause a diuresis of phosphates This cannot be accomplished and the phosphates are not excreted in the urine In renal rickets there should be a normal or slightly lowered serum calcium and a high serum phosphorus The serum calcium would be lower and the serum phosphorus higher if the compensatory hyperplasia of the parathyroids was not present Mitchell believes that a second mechanism exists Since the damaged kidneys cannot excrete phosphorus there is excretion through the bowel The increase of phosphorus in the bowel interferes with calcium absorption as it fixes the food calcium in an insoluble phos

"Fanconi's syndrome" is also applied to a condition occurring in young people and characterized by (1) a severe, progressive refractory congenital hypoplastic anemia with pancytopenia, there being neutropenia and thrombocytopenia, (2) hypoplasia of the bone marrow, (3) a generalized brown melanin like pigment throughout the skin due to hemosiderosis and (4) various associated congenital anomalies which comprise microcephaly, hypogenitalism and other genito urinary abnormalities, strabismus, unilateral deafness, syndactylia, congenital heart disease, and bone abnormalities affecting particularly the forearms and thumbs. Confusion is best avoided by using the term de Toni Fanconi syndrome for the metabolic disorder under consideration and applying the name Fanconi's anemia to the other disease which is of particular interest to the hematologist.

Nephrocalcinosis

Nephrocalcinosis is defined as the deposition of calcium in the renal parenchyma and may involve the pyramids, the cortex or both. It is a general term used to describe several conditions of widely differing etiology. Normally the calcium phosphorus and protein of the blood exist in intimate relationship with each other. Approximately 60 per cent of the serum calcium is in combination with protein and is not diffusible through a colloidal membrane. The concentration of calcium and phosphorus varies with the pH in the blood. As the calcium level rises to 12 mg per 100 cc or more, significant percentages of phosphate become non diffusible. Normally, approximately 70 per cent or more of ingested calcium is excreted by way of the bowels while the greater part of the phosphorus is found in the urine. The renal thresholds for calcium reabsorption is at the blood level of 7 mg per 100 cc. Therefore this ion is continuously being lost in the urine and the rate of loss is roughly proportional to the height above 7 mg per 100 cc of the blood calcium level. The renal excretion of phosphorus is intimately related to the activity of the parathyroid gland. In parathyroid hyperactivity there is increased phosphaturia while hypoactivity results in lessened excretion. The calcium output is secondarily affected in both instances. With rapid excretion of phosphates, the blood phosphates become subnormal, producing higher calcium levels and consequently increased calcariuria. Conversely when the excretion of phosphate is less than normal, the blood level increases and the blood calcium level falls with consequent decrease in the amounts excreted in the urine. Therefore, with rapid excretion of phosphorus, there is increased excretion of calcium and vice versa. An exception to this is in uremia in which condition low output of phosphorus by the decreased glomerular mass may be associated with a rapid loss of calcium because of the associated acidosis. This is most probably due to the inability of the tubules to elaborate enough ammonia to equalize the acid products of metabolism and therefore a fixed base is called upon instead.

Although this condition has been known for several years having first been described by Butler, Wilson and Farber in 1936, Albright coined the name "nephrocalcinosis" in 1940. The condition may occur in association with stones of the kidneys or as a solitary manifestation. In nephrocalcinosis two important functions are lost: the ability to make ammonia and to excrete an acid urine. Both of these mechanisms tend to conserve base. In consequence calcium is excreted in the urine in increased amounts

association with these changes The epiphyseal separation comprises a subepiphyseal separation with fracture of the type commonly seen in scurvy The cortical erosions involve the upper medial borders of the tibias and the distal portions of the shafts of the ulnas and other bones Periosteal elevation and thickening may occur

An important manifestation is retardation of skeletal maturation The development of the bones of the carpus may lag one or two years behind the chronological age This does not occur in ordinary rickets It appears that the epiphyses and the centers of the bones are not slow in developing but rather are so demineralized that the bones are not visible Delay in the development of the bones is not pathognomonic of renal rickets as it occurs in hypofunction of the pituitary and thyroid, in long standing constitutional diseases, and other osteonephropathies Cyst formations in the metacarpals are common Bilaterally symmetrical insufficiency fractures and notching of the head of the radius may also occur The cortex of the long bones is thinned, the diaphyseal ends of the bones are broad, the epiphyseal lines are widened and irregular, and there is bowing The bones may show stippling Periosteal bone formation may be present Numerous fractures may occur The appearance of the skull is variable The bones may be decalcified and show cystlike patterns and mottling due to irregular areas of decalcification In other instances, the appearance is more that of Paget's disease, homogeneous density or multiple areas of increased density with fuzzy borders being distributed irregularly throughout the bones of the skull Prominent bosses may occur in the parietal and occipital regions The inner table of the skull is sharply defined, the outer table is irregular and poorly outlined The roentgen changes may be so similar to those of hyperparathyroidism syphilis, or chronic osteomyelitis that differentiation may be made only by determining the presence of chronic renal disease and after study of the chemistry of the blood Many of the patients show diminished kidney function and intravenous pyelographic study proves unsatisfactory It is important to carry out retrograde pyelographic studies in order to determine the site and extent of obstructive lesions There frequently develops metastatic calcification The common locations are the blood vessels the kidneys the subcutaneous tissues and the skin Calcific depositions may occur in the lungs and other organs of the body, practically every organ having been involved in the reported cases Calcification in the costal cartilages is common An instance has been reported in which the peripheral arteries were extensively calcified in a child of five years Metastatic calcification is more common than in primary hyperparathyroidism It also occurs in hypervitaminosis D, congenital syphilis periarteritis nodosa leukemia neoplasms, infections of bone and osteogenesis imperfecta hence is not pathognomonic of renal rickets

Fanconi's Syndrome De Toni-Fanconi's Syndrome

Fanconi s syndrome is a form of osteomalacia associated with excessive renal loss of amino acids glucose bicarbonate, phosphorus, potassium and peptides Acidosis eventually results in the formation of renal calculi with changes in the tubules and glomeruli, cirrhosis of the liver or both The patients show dwarfism and severe rachitic changes The blood chemistry is altered and there is a low level of serum phosphorus There is much confusion in the literature due to the fact that the name

roentgen manifestations are similar to those in renal rickets, the laboratory findings in nephrocalcinosis are entirely different and establish the diagnosis There is no elevation of the non protein nitrogen or blood urea nitrogen Anemia is absent or mild The blood phosphorus is low and the serum calcium is normal or slightly low There is a chloride acidosis with elevation of the serum chloride and lowering of the carbon dioxide combining power The important feature is an increase in the excretion of calcium in the urine The Sulkowitch test is strongly positive as a rule The calcific deposits in nephrocalcinosis are extensive mottled, finely granular densities The renal calcifications are distributed throughout the cortex and vary from 1 to 2 mm or more in diameter There are usually no calculi in the renal elements, ureters or bladder and urographic studies as a rule demonstrate normal renal pelves and calyces through the overlying calcifications There is usually no osteoporosis The heart shadow is not enlarged and the lung fields are normal Extensive calcifications of the vascular structures may occur

Differential Diagnosis In nephrocalcinosis there is no retention of nitrogenous products or phosphorus in the blood It differs from the Fanconi syndrome in three respects (*a*) there is no glycosuria or abnormal organic acid excretion in the urine (*b*) there is often an associated hyperchloremia (*c*) there are calcifications in the renal parenchyma the so called nephrocalcinosis The picture is different from that in hyper parathyroidism which is characterized by multiple calculi in the renal pelves and calyces The roentgen appearance is similar to the metastatic calcification which may occur in advanced hyperparathyroidism The difference in the osseous changes in the two diseases is of importance in differential diagnosis Calcification due to tuberculosis is more amorphous tends to be grossly lobulated and is usually unilateral Calcific deposits in renal tumors vascular anomalies infarcts and similar conditions are differentiated by the uniformity of the calcific deposits in calcinosis

Idiopathic Hypercalcariuria

The fourth entity which gives rise to rickets is termed idiopathic hypercalcariuria The pathogenesis of the rickets is the same as in vitamin D deficiency However instead of there being a decrease in the calcium absorption, there is an increase in the calcium excretion The renal threshold for calcium is low with resultant excessive excretion of calcium by the kidney even though the level of calcium in the serum remains normal Bone changes and renal calculi ensue

It is essential to understand that the group of four closely related but entirely distinct syndromes characterized by bone changes associated with renal disease renal rickets, Fanconi s syndrome nephrocalcinosis and idiopathic hypercalcariuria, comprise the conditions grouped under the heading of "osteonephropathy The bone changes comprise a combination of rickets plus hyperparathyroidism and one or the other may predominate

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with a resultant fall in the serum calcium level This leads to a parathyroid hyperplasia which elevates the serum calcium level A phosphate diuresis may occur since the glomerular function is not impaired The hyperphosphaturia leads to hypophosphatemia In consequence there is a normal or slightly lowered serum calcium level and a low serum phosphorus level The calcium cannot be deposited in osteoid, consequently



FIG 208 Nephrocalcinosis A There are multiple small irregular areas of calcific density distributed irregularly throughout the parenchyma of the kidneys B The renal elements are opacified and demonstrate that the calcific depositions do not lie in the renal pelvis or calyces

rickets develops In nephrocalcinosis there is a hypophosphatemic type of rickets similar to infantile rickets Important concomitants of the syndrome associated with the hypercalcariuria are nephrocalcinosis and nephrolithiasis When the calcifications become sufficiently marked damage of the glomeruli occurs

The affected individuals are dwarfed or stunted in growth There is polyuria and bone deformities due to rickets While the clinical and

CALCIUM, PHOSPHORUS AND PHOSPHATASE AS AIDS IN THE DIAGNOSIS OF BONE LESIONS

In many instances, disease of the bones may be recognized and diagnosed with accuracy solely by x-ray study. In certain cases, however, correlation of all the available clinical data, particularly the laboratory criteria, is necessary for final evaluation. The serum calcium, phosphorus, and phosphatase values vary with age. The normal serum calcium averages 9.5 to 11.0 mgm. per 100 cc. for adults and 10.0 to 12.0 mgm. for infants and children. The serum phosphorus ranges from 2.5 to 4.5 mgm. per 100 cc. for adults and 4.0 to 6.5 mgm. per 100 cc. for children and infants. The determinations must be made on serum which is free of hemolysis. The serum calcium is composed of two fractions: diffusible calcium which is practically completely ionized and non-diffusible calcium. The latter comprises about 45 per cent of the total and is bound to protein, hence is dependent on the amount of albumin. In the absence of renal insufficiency or hyperglobulinemia, one gram of protein binds approximately 0.5 mgm. of calcium and a relatively slight decrease of serum protein in nephrosis or malnutrition results in a very marked lowering of the calcium level. Failure of recognition of the important role played by protein may obscure an actual hypercalcemia in hyperparathyroidism or in other instances lead to the false assumption that hypocalcemia is present.

Phosphatase. The phosphatase activity of serum has been extensively studied in numerous diseases. On the basis of its activity at different pH levels, phosphatase may be divided into four types: (1) an alkaline type with optimal activity at about pH 9.3 occurs in the bone, ossifying cartilage, intestine, kidney, mammary gland, lung, spleen, blood serum, leukocytes and adrenal cortex, (2) a phosphatase with optimum activity at pH 6.0 is present in mammalian erythrocytes, (3) a phosphatase with optimum activity at pH 5.0 is found in the spleen, liver, pancreas, kidney, prostate, and blood serum, and (4) a phosphatase in certain yeasts with optimal activity at pH 3.0 to 4.0. From the clinical point of view, the alkaline phosphatase at pH 8.6 to 9.3 and the acid phosphatase at pH 5.0 are important. The erythrocyte phosphatase is significant only in that hemolysis-free serum should be used for the determination of both phosphatases, particularly the acid type. Numerous methods for determining the phosphatase activity are in use, each utilizing a different set of standards. The units differ because of differences in the substrate which is utilized. In the Bodansky method, the phosphorus liberated during the incubation of serum with beta glycerophosphate is determined. This is one of the most widely used methods. In the case of alkaline phosphatase, the normal values are 0.2 to 1.0 Bodansky units and 0.6 to 3.0 King-Armstrong units.

The diseases in which the alkaline phosphatase is elevated with sufficient frequency to be of diagnostic significance comprise: rickets, osteomalacia, Paget's disease of bone, renal rickets or renal osteodystrophy, hyperparathyroidism with osteitis fibrosa cystica generalisata, neurofibromatosis of bone, osteogenic sarcoma, metastatic carcinoma of bone, Hodgkin's disease and other lymphoblastomas which involve bone, Boeck's sarcoid, carcinoma of the prostate with metastases, jaundice of the obstructive type, extensive fractures in the healing phase, and the last trimester of

HYPOPHOSPHATASIA

Rathbun reports what he believes to be the first account of faulty bone development associated with absence of alkaline serum phosphatase The patient was a poorly nourished infant with deformities of the wrist and bowing of the legs Only four rounded plaques of bone were palpable in the frontal and parietal regions of the skull, the vault was otherwise soft and felt like a balloon filled with water There was beading at the costo chondral junctions The blood showed slightly elevated calcium and phosphorus Except for a markedly diminished excretion of calcium and phosphorus the urinary studies were negative Repeated determinations showed complete absence of alkaline serum phosphatase or an extremely low figure The roentgen findings comprised marked decalcification throughout the skeleton, deformities of the ribs and lack of normal calcium density at the metaphyseal ends of the long bones The epiphyses were unaffected Fractures of the metaphyses of the radius and ulna were noted The histopathologic findings at postmortem were most unusual The kidneys showed the tubules distended with casts The picture at the growing ends of the bones resembled that in rickets The vault of the skull showed a normal framework of osteoid tissue but no deposition of calcium Samples of various tissues were found to be abnormally low in phosphatase The condition was considered to be a primary failure by the osteoblasts to produce alkaline phosphatase and represented a mesenchymal differentiation defect

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THE CELIAC SYNDROME

The celiac syndrome is characterized by a failure of the small intestine to absorb fat carbohydrate calcium and in some instances phosphorus and occurs in the disease of childhood known as Gee s Herter s or celiac disease It may also develop in steatorrhea associated with lympho sarcoma of the intestines or mesenteric lymph glands and gastrocolic fistula There is usually persistent diarrhea the stools being bulky foul fatty and soapy The disease is characterized by progressive wasting of the body tissues There is severe calcium loss due to combination of the calcium with the unabsorbed fat in the intestine This causes irrita bility tetany and spasmophilia due to changes in the nervous system Skeletal anomalies ensue with marked osteoporosis deformity and dwarf ism Opacities in the lens of the eye and disturbance in the blood clotting mechanism may also develop Anemia tongue lesions dermatoses and disturbance of the water metabolism are of less common occurrence

The roentgen changes in the bones comprise osteoporosis spontaneous fractures deformities and delay in union of the epiphyses Striations are present in the bones and are indicative of increased calcium deposition in the metaphysis adjacent to the epiphysis Dwarfism is common When bone growth is not inhibited the patient grows normally and the characteristic manifestations of rickets may develop

Calcium occurs in the blood in organic compounds as indiffusible calcium and in inorganic salts as diffusible calcium The former constitutes slightly less than half and the latter slightly more than 50 per cent of the blood calcium The diffusible blood calcium is practically completely ionized The total content of calcium in the blood is 10 to 11 mg per 100 cc The functions of calcium are multiple In the form of phosphate and carbonate it constitutes the principal content of bone dentin cementum, and enamel It is present in all the tissues and the glandular secretions Two functions of calcium are particularly important It is essential for the coagulation or clotting of the blood and the maintenance of the excitability of the nerves, particularly the motor nerve endings As calcium is readily available deficiencies occur only in consequence of severe malnutrition after famines or among the grossly underprivileged In most instances calcium deficiency is caused by lack of absorption There are two consequences of calcium deficiency as regards the skeleton The preparatory calcification of the cartilage during the growth period and the calcification of newly formed organic bone matrix are prevented or grossly disturbed Resorption of bone in order to mobilize calcium becomes increased with the development of osteoporosis Rickets or osteomalacia ensues the age of the calcium deficient individual being the determining factor as to which disease develops

Bone is a storehouse for calcium This view is justified by the fact that calcium is mobilized by resorption of bone The skeleton should more properly be considered a source of calcium during an emergency Because of rapid excretion excessive intake of calcium in normal individuals results merely in a temporary elevation of the blood calcium level Hypercalcemia of long duration occurs in primary or secondary hyperparathyroidism hypervitaminosis D and extensive destruction of bone by tumors In the presence of prolonged hypercalcemia depositions of calcium salts as phosphates and carbonates take place in many organs particularly the kidneys the arteries the heart, the lungs and the mucous membranes This calcification has been termed *metastatic*, although the term is erroneous The calcification occurs especially in organs and tissues which are rich in phosphates It is essential not to confuse "metastatic calcification with the *dystrophic* form The latter comprises calcification of degenerated or necrotic tissue and is due to local elevation of the hydrogen ion concentration because of a reduction in carbon dioxide tension Dystrophic calcification is independent of the local phosphatase concentration

Calcium Excretion Extensive and careful calcium balance studies are essential in order to determine whether there has been a net gain or loss of calcium during a given period The relationship between the calcium in the urine and the stools is of great significance Normally, about two thirds or more of the calcium is excreted in the feces and one third or less in the urine This ratio changes markedly in diseases of the parathyroid gland In hyperparathyroidism a greater percentage is excreted in the urine with a resultant negative calcium balance The ratio shifts in all hypocalcemic states and a greater proportion or all of the calcium is excreted in the feces Determination of the quantity of urine calcium has proven of great importance A normal individual excretes less than 100 mg per day In hyperparathyroidism and hyperthyroidism the excretion is greatly increased and may rise to more than 200 mg daily The excretion increases in diseases associated with rapid decalcification of a portion of or the entire skeleton

pregnancy Diseases in which the phosphatase levels are abnormally low are cretinism and scurvy With the exception of obstructive jaundice or liver damage, it appears that increased phosphatase activity depends on osteoblastic activity In Paget s disease of bone, the osteoblastic activity predominates over the osteoclastic activity and the phosphatase is uniformly elevated when large areas of bone are affected

In hyperparathyroidism, the elevation of the phosphatase level corresponds to the extent of involvement of the bone and is correlated with the degree of decalcification as manifested on roentgen examination The phosphatase is increased in rickets and osteomalacia in the stage of bone repair with abundant osteoid tissue Multiple myeloma is characterized by destruction of bone with little or no attempt at repair and in this disease phosphatase is usually normal Therefore it appears that in the absence of jaundice phosphatase activity depends on osteoblastic activity Since the serum phosphatase is elevated in a large variety of conditions this finding cannot be considered pathognomonic of any single disease In correlation with the roentgen findings, it is of great significance The acid phosphatase is particularly important since the relationship is limited to metastases from carcinoma of the prostate

Carcinoma of the prostate with metastases to the skeleton is generally characterized by a high serum acid phosphatase elevated values for serum acid phosphatase being present in about 85 per cent of the cases Prostatic tissue has a high acid phosphatase content The same is true of carninoma of the prostate unless the neoplasm is very undifferentiated The effectiveness of castration or estrogen therapy is striking and there is a marked change in the acid phosphatase as improvement occurs The level of the acid phosphatase in the blood becomes markedly elevated only in metastasizing carcinoma of the prostate Slight elevations may occur in patients with an altered alkaline phosphatase level in Paget s disease hyperparathyroidism and metastatic carcinoma from the breast

Calcium Calcium is indispensable for the development and function of the animal body Man requires a daily intake of about 0 8 grams for an average weight of 70 kilograms During growth pregnancy and lactation greater amounts of calcium are necessary Children up to the age of twelve years need about 1 gram per day During the years from twelve to twenty the daily allowance should be 1 4 grams for boys and a little less for girls The intake during pregnancy should be 1 5 grams and during lactation 2 0 grams Of the calcium in the human body 99 per cent is in the skeleton and the teeth with only about 1 per cent in the blood and other tissues The absorption of calcium from the gastrointestinal tract is regulated by vitamin D and occurs mainly in the upper part of the small intestine An important role is played by the pH of the contents of the intestinal tract Most calcium salts are insoluble in alkaline fluids In consequence acidity of the intestinal contents is a prerequisite for normal absorption of calcium The acidity of the small intestines is increased by the intake of fats Oxalic acid contained in spinach forms insoluble calcium salts and prevents calcium absorption Phytic acid which is present in whole wheat acts in a similar manner Calcium may combine with fatty acids to form soaps if the production of bile or pancreatic juice is disturbed with consequent lessening of the absorption of calcium fats and vitamin D Vitamin D deficiency aggravates the calcium deficiency and a vicious circle is established

renal insufficiency and renal calculus formation do not occur in association with hyperthyroidism in spite of the increased excretion of calcium and phosphorus in the urine Metastasis to the skeleton from carcinoma of the prostate occupies a unique place in bone diseases because of the increased serum acid phosphatase activity in the majority of cases

Phosphorus Phosphorus is a basic constituent of bone and teeth and serves as an intermediary compound of carbohydrate metabolism It is indispensable to the normal function of the human body The daily requirement for a normal adult is 1 3 mgm In children the need is slightly less while in pregnancy and lactation it is increased Phosphorus is so abundant in ordinary foods that extrinsic deficiency is very unusual Its absorption from the intestinal tract is dependent in part on calcium With absence of fat in the diet calcium phosphate which is not readily soluble is formed and the absorption of phosphorus is retarded the greater portion being eliminated in the stool The presence of fat in the diet decreases the excess of calcium by the formation of insoluble soaps and indirectly increases the absorption of phosphorus About two thirds of the absorbed phosphorus is excreted in the urine and the remainder is eliminated with the feces Vitamin D furthers calcium absorption from the intestinal tract and indirectly affects the absorption of phosphorus Its excretion by the kidneys is increased by the action of the parathyroid hormone as the hormone serves to decrease the reabsorption of phosphorus from the glomerular filtrate in the tubules

The amount of phosphorus in the human blood is 4 to 5 mg per 100 cc It is present in the blood in almost equal proportions in the inorganic ionized and organic non ionized forms The organic portion is found almost entirely in the red blood cells while the inorganic phosphorus is in the serum Eighty five per cent of the total phosphorus content of the body is contained in the skeleton and the teeth

Intoxication by elementary or metallic phosphorus is practically unknown today It was common in the manufacture of matches in the days prior to the utilization of the inactive red variety and was due to the use of the highly toxic yellow phosphorus The presence of osteosclerosis in patients suffering from phosphorus poisoning led to the use of this element in therapeutic doses in rickets or after fractures on the assumption that bone formation and calcification would be promoted Metallic phosphorus in combination with cod liver oil was used in the treatment of rickets Since the discovery of vitamin D this has been discontinued

After the ingestion of phosphorus sclerotic bands develop in the bones This has been utilized to determine the rate of apposition of bone in the metaphysis and the epiphysis, the bands indicating the bone which was formed during the period of phosphorus medication Microscopic and histologic investigations indicate that osteosclerosis of growing bone is due to failure of resorption Disturbance of resorption of the calcified cartilage results in the persistence of thick trabeculae of cartilage which later form the core of markedly thickened bone trabeculae The trabeculae in the metaphysis escape resorption for long periods of time and occupy a greater portion of the bony shaft with decrease in the size of the marrow cavity

The reasons for the toxic effects of ingested metallic phosphorus on bone and cartilage are not understood It is believed that it affects the ground substance of the calcified cartilage and bone The intercellular substance acts as a foreign body as evidenced by the presence of numerous

In primary hyperparathyroidism the amount of calcium in the blood is usually greater than normal and the phosphorus is low With renal insufficiency of sufficient degree to cause retention of blood urea nitrogen, the phosphorus level is normal or elevated and the calcium level is normal More calcium is excreted in the urine than normally except in association with severe renal insufficiency Hyperparathyroidism is frequently an obscure disease and its recognition in many instances is dependent upon careful correlation of the x ray and laboratory studies The bone lesions of this disease have received great stress Less well emphasized is the serious renal damage which ensues in untreated cases of hyperparathyroidism Renal stones occur frequently and may cause hydronephrosis pyelonephritis, and other complications The parenchyma of the kidney may become calcified and cause severe, irreversible renal insufficiency and uremia In multiple myeloma the serum calcium may be increased The phosphorus levels are unchanged and the serum phosphatase is normal in practically all cases There is usually increased excretion of calcium in the urine Senile and postmenopausal osteoporosis are characterized by normal serum calcium phosphorus and phosphatase values It appears that there must have been a negative calcium balance of long duration in order for the bone to have become so markedly decalcified Rarely, the serum calcium phosphorus and phosphatase are elevated

It is not always possible to establish an absolutely definite diagnosis solely on the basis of the blood chemistry studies However certain important facts can be established The calcium and phosphorus levels are reciprocally related in most instances Hence with a high calcium level the phosphorus is low and vice versa With calcium, phosphorus and vitamin deprivation as in rickets and steatorrhea both calcium and phosphorus may be low or normal In hyperparathyroidism there is diffuse, generalized bone disease and x ray studies of any portion of the skeleton reveal definite changes Paget s disease on the contrary is never generalized and roentgen study reveals normal bone in certain portions of the skeleton In Paget's disease one portion of the bone may be normal while another part shows changes which are typical of osteitis deformans Metastatic carcinomatosis is apt to be confused with multiple myeloma or hyperparathyroidism The discovery of a primary tumor establishes the diagnosis The biochemical studies in multiple myeloma and carcinomatosis are so closely allied that differentiation cannot be made on these grounds alone The roentgen manifestations are bizarre There are areas of pigmentation in the skin and precocious puberty in the female The biochemical changes may resemble those of mild hyperparathyroidism the alkaline phosphatase particularly tending to be elevated

Secondary hyperparathyroidism such as is found in renal insufficiency may lead to osteitis fibrosa cystica generalisata identical with the primary disease The biochemical findings are the important differential feature, particularly the elevated phosphorus and low or normal calcium When renal insufficiency is due to primary hyperparathyroidism the characteristic high calcium and low phosphorus are not present and there may be normal levels of calcium and phosphorus with resultant confusion in the differentiation of the conditions Chronic hyperthyroidism causes osteoporosis with increased secretion of calcium and phosphorus There usually are normal levels of serum calcium and phosphorus The possibility that hyperthyroidism is present should be borne in mind in all patients with marked presenile osteoporosis It is important to stress that

FIBROUS DYSPLASIA

Introduction The first case of fibrous dysplasia of bone was described by Weiland in 1922 Lichtenstein in 1938 reported a series of cases and was the first to employ the term "fibrous dysplasia of bone" It is of relatively common occurrence and with the increasing use of roentgen methods of diagnosis larger numbers of cases will doubtless be diagnosed in the future Clinical diagnosis is not possible, the establishment of the diagnosis being dependent on roentgen or histopathologic methods of study The condition has been known in the literature by various names the most common of which are fibrocystic disease osteitis fibrosa cystica osteodystrophia fibrosa, fibroma of bone, regional fibrocystic disease disseminated osteodystrophy, focal osteitis fibrosa, fibro osteoma of bone

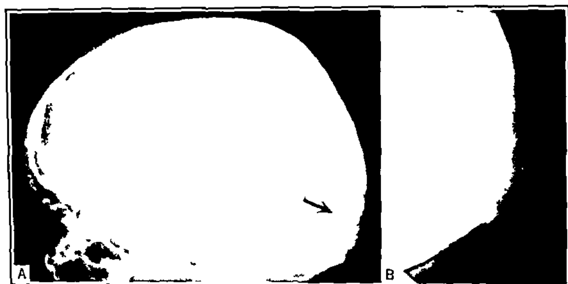


FIG 209 Fibrous Dysplasia of Skull *A* Lateral view of the skull *B* Spot film of the occipital region There is a localized area of increased radiance in the occipital region The tables of the skull are thinned and expanded and there is granular mottling in the affected area The roentgen diagnosis of fibrous dysplasia was confirmed by histopathologic study

polyostotic fibrous dysplasia and others It occurs in two forms the monostotic for which the name fibrous dysplasia of bone is now universally used and the polyostotic variety which is also known as Albright's disease In the former there is a solitary osseous lesion The polyostotic form is characterized by the presence of multiple bone lesions occurring in association with areas of abnormal pigmentation in the skin and precocious puberty in the female

Etiology The etiology of fibrous dysplasia of bone is unknown It has been postulated that endocrine imbalance plays an important role in the polyostotic forms which are associated with extraskeletal anomalies Many observers are of the opinion that the two varieties of the disease merely represent different expressions of the same basic condition The role of injury in the etiology has been stressed by Schlumberger He believes that trauma is an important etiologic factor in the monostotic variety and that this form of the disease has nothing in common with polyostotic fibrous dysplasia despite the fact that the lesions in the two

osteoclasts about the bone which form after the ingestion of phosphorus Simultaneously the calcified cartilage of bone becomes unresorbable The reaction of the surrounding tissue to the resorption fast foreign body consists of the apposition of new bone along the surface in an apparent attempt to encapsulate the damaged tissue The damage to the matrix and the resultant over production of bone continue as long as the excessive intake of phosphorus persists The osseous changes of phosphorus intoxication are not specific Other metallic poisons such as lead and arsenic result in the development in the metaphyseal and epiphyseal growth zones of osteosclerotic bands which are similar to the phosphorus bands In cases of metallic poisoning, the skeleton contains large amounts of the toxic agent In later years these are gradually mobilized and there is slow resorption of bone The delayed reaction accounts for the persistence of clinical symptoms of metallic poisoning long after the ingestion of the poisonous substance has ceased

a small percentage of the patients but the significance of this manifestation is not clear nor is it known whether it is merely coincidental. The lesions of fibrous dysplasia may progress slowly or remain stationary for many years. Healing usually follows removal of the fibrous tissue although recurrences have been reported. Sarcoma has occurred in patients with fibrous dysplasia but it is generally believed that while the two diseases may coexist, they do not merge into each other. During the period of skeletal growth there is a tendency to gradual progression with slight increase in the size of the lesion. During adult life the condition usually remains stationary.

In females there are skeletal manifestations of polyostotic fibrous dysplasia concurrent with precocious puberty and anomalous pigmentation of the skin, a triad known as Albright's syndrome. The bone changes



FIG 211 Fibrous Dysplasia. There is an area of increased radiance involving the ilium in the region above the acetabulum. This area appears cystic and presents irregular margins. The lesion was discovered during an x ray study after a trauma. The roentgen diagnosis of fibrous dysplasia was confirmed by histopathologic study of a specimen removed at operation.

may occur alone and are considered the central point of the syndrome. The condition appears to originate in bone with normally developed cortex and which has had a normal premorbid development. It appears unlikely that, though starting in childhood, it can be congenital in origin. In a postmortem study by Stenbergh and Jones there were hypertrophy of the basophil apparatus of the hypophysis with microscopic adenomas, atrophy of the suprarenal cortex, relative hypertrophy of the suprarenal medulla, hyperplasia of the thyroid, and no central nervous system lesions. It usually subsides at puberty. The site of predilection is the long bones, the tibia and femur being most often affected. It may be monomelic or bimelic, unilateral bimelic involvement being more frequent. Bilateral involvement is found in pronounced cases and lesions occur in the long bones, ribs, skull and pelvis, and rarely the vertebræ. In the long bones the metaphyses and diaphyses are affected. The epiphyses are not primarily involved but may show involvement if the process continues to be active after growth has been completed.

varieties of the disease show little or no demonstrable differences. The monostotic form is much more common. The age of the patients in the previously recorded cases has varied from a few months to over sixty years. It is believed that practically all cases originate in childhood although the exact date of onset frequently is indeterminate. Cases have been reported in all races. There is no evidence of a familial or hereditary tendency. The disease has been found in many of the bones of the body, the common sites being the femur, tibia, fibula, radius, humerus, metacarpals, phalanges, skull and ribs. It has also been found in the maxilla, mandible, vertebra, and scapula. The condition has been considered a disturbance of the normal reparative processes subsequent to traumatism to bone. Lichtenstein and Jaffe are of the opinion that the lesion is a manifestation of a congenital defect characterized by perverted activity of the bone producing mesenchyme.

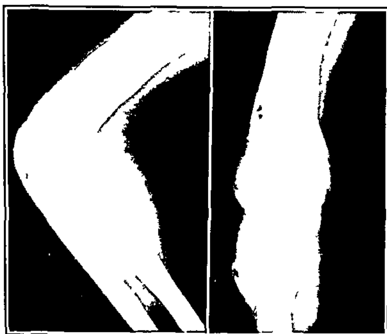


Fig. 210. Fibrous Dysplasia. The bones about the elbow show expansion, cortical thinning and irregular granular mottling, the characteristic manifestations of fibrous dysplasia.

Clinical Manifestations. The clinical history is extremely variable. Not infrequently symptoms are entirely absent. In many instances, the onset is related to a specific trauma, while in a great number of others there is a gradual onset with no antecedent injury. There may be a long standing history of pain, tenderness and swelling in the region of the lesion. Persistent mild pain is common. A localized asymptomatic swelling or a pathologic fracture may be the first manifestation. Fever, leukocytosis, and elevation of the sedimentation rate do not occur. The serum calcium and phosphatase levels may be slightly elevated; otherwise blood chemistry studies reveal no abnormalities and the Wassermann test is negative. Calcium and phosphorus excretion are normal. Pigmentation of the skin and precocious puberty accompany the polyostotic form of the disease in females, the menarche occurring at the age of ten to twelve years. A tumor has not been described in any instance. Premature skeletal growth may occur in the polyostotic form. Hyperthyroidism has been noted in

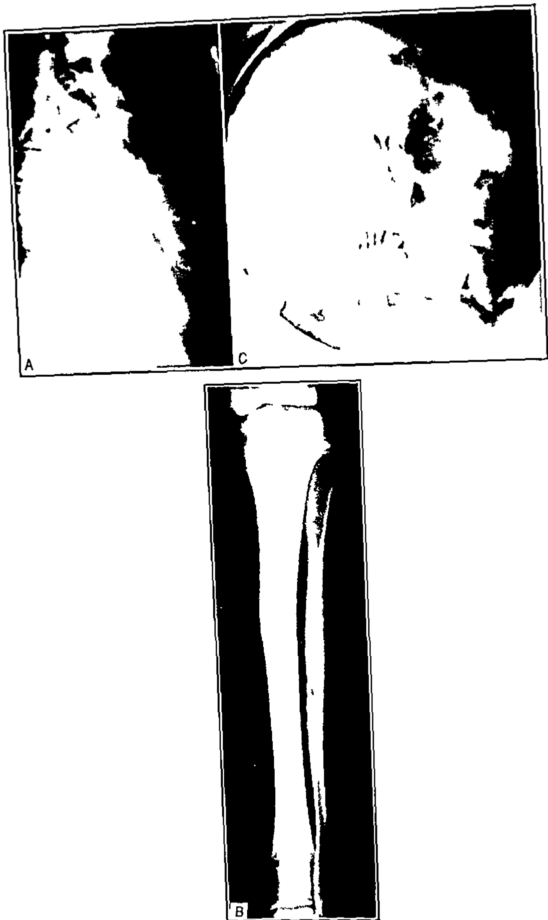


FIG 212 Fibrous Dysplasia *A* The ribs There are multiple deformations with cystic rarefactions involving the left ribs *B* The right lower leg The bones of the leg show granularity irregular rarefaction and cortical thinning *C* The bones of the skull and face show increased density and asymmetry The patient is a male fifty three years of age Changes similar to those illustrated were distributed throughout practically all of the osseous structures Despite the widespread extent of the lesions there were no symptoms referrable to the bones the condition being discovered incidentally on roentgen examination The roentgen diagnosis of fibrous dysplasia was confirmed by histopatholgic study of a fragment of bone from the tibia

Roentgen Manifestations The earliest sign is the presence of a minute, rounded area of increased radiance within the cortex of the bone In the monostotic form, there is a solitary, rounded defect, while in the polyostotic variety there are multiple lesions which are distributed unilaterally or bilaterally The character of the changes is similar in both forms of the disease The localized radiance in the bone is due to replacement of the osseous structure by fibrous tissue and miniature spicules of bone producing a homogeneous change with absence of trabecular structure and bony architecture which is best characterized as a ground glass appearance The affected area is usually sharply defined and clearly demarcated from the adjacent normal bone The defect involves both cortex and medulla and is usually situated eccentrically The bone may be expanded to a slight or a marked degree Periosteal reaction is usually absent The overlying soft tissues are not affected and the uninvolved bone is of normal density

As the lesions increase in size pressure is exerted on the adjacent bone The growth of the process is along the lines of least resistance and defects in the medulla usually extend toward the medullary canal and present a crescentic margin Lesions situated near the outer aspect of the cortex may produce expansions or rounded elevations of the surface of the bone Spontaneous fractures may ensue but heal readily with new bone formation There is no periosteal elevation or thickening except as manifestations of the healing of a fracture Osteoporosis does not occur and there is no softening of bone or deformities Localized bone sclerosis may be present at the margins of the lesion and is a manifestation of resistance to the advancement of the process *In cancellous bone, there is little resistance* to the progress of the disease and slight or no sclerosis whereas in the petrous portion of the temporal bone and the base of the skull extensive sclerosis ensues The roentgen picture varies widely being dependent on the distribution of fibrous tissue in the bones and the extent of the metaplastic process The changes are more apt to progress during the active period of growth than after adolescence when the bones have ceased to undergo increase in size

The disease has been reported as occurring in practically every bone in the body The bone lesions are located in the diaphysis or the metaphysis the epiphysis apparently never being the primary site A juxtaepiphyseal lesion in the metaphysis may extend through a closed epiphyseal line The area of involvement is usually sharply demarcated with clearly defined margins Small lesions tend to show a narrow band of increased density at the edge of the lesion Longitudinal bone growth is not affected in most instances although very extensive lesions may be associated with an initial acceleration of growth followed by premature closure of the epiphysis

In some instances the area of radiance presents a multilocular appearance This is due to the presence of coarse columns or trabeculæ of bone protruding into the lesion from its periphery In multiple lesions which have undergone coalescence there is irregular mottled density The areas in which calcification is absent show the density of soft tissue Other areas appear dense eburnated and loculated due to overlapping of the lesions and the healing of pathologic fractures A zone of increased density may surround the area of fibrosis This zone may be prolonged at one point in a tongue like projection which has been likened to candle flame Repeated fractures or bowing of the

tains multiple trabeculae of partly calcified new bone formed by direct metaplasia of the connective tissue. A few large osteoclasts are usually present. The affected bone is firm, rubbery, yellowish white in color and fibrous in character. The abnormal fibrous tissue has a gritty sensation due to the presence of multiple immature partially calcified bone trabeculae. A cyst containing fluid may rarely be found. The areas of new bone formation may range from acidophilic and osteoid tissue to adult calcified bone. Giant cells may be present but they can be distinguished from those seen in giant cell tumors by the fact that the nuclei range from 2 to 10 in number rather than from 15 to 200. The fibrous process may appear to extend into the surrounding muscle in the absence of a fracture demonstrable on the roentgenogram. This is particularly apt to be the case if the cortex is very thin.

Prognosis and Treatment. The prognosis is good. The condition is self limited. It is generally accepted that malignant degeneration does not ensue, although isolated cases have been reported in which neoplastic changes have occurred in areas of fibrous dysplasia. Simple excision or curettage and bone grafting usually result in cure. Pathologic fracture is not uncommon and is an indication for surgical intervention.

Differential Diagnosis. In differential diagnosis it is necessary to consider a large number of lesions. In non osteogenic fibroma the roentgen appearance is very similar to that of fibrous dysplasia. The former does not present a ground glass appearance roentgenographically. In Brodie's abscess there is absence of trabeculation with little or no cortical expansion, although sequestrum may be present. Hyperparathyroidism can be differentiated by the altered chemical changes in the blood. The bone changes are somewhat similar to those of generalized osteitis fibrosa cystica. In the developing stage, however, there is one important difference. In generalized osteitis fibrosa cystica there is extreme osteoporosis which results in an extreme degree of deformity from bending of the affected bones. In fibrous dysplasia deformity is rare and is due to enlargement of the individual bones. While the bones become weak from thinning of the cortex and frequently fracture, bending does not occur. There is no periosteal reaction in fibrous dysplasia except in the presence of fractures and the expanded areas in the cortical and subcortical bones are not true cysts but are filled with fibrous tissue and are sharply delineated by the adjacent bony structure. The intervening bone is normal and there is no osteoporosis. In osteitis fibrosa cystica the vacuolated areas are true cysts in the bone which arise in the cortical structures. They may have definite lining membranes of fine fibrous tissue and contain clear straw colored or blood tinged fluid. The remainder of the bony structure shows marked osteoporosis. Usually the clinical findings and the mineral metabolism establish the diagnosis although in some instances biopsy is necessary. Solitary bone cyst occurs in individuals under twenty years of age, involves the diaphyseal ends of the long bones and is in close relation to the epiphyseal plate. The lesion is central in origin and is accompanied by true symmetrical expansion of the shaft of the bone.

Giant cell tumor usually occurs in individuals over twenty years of age. The lesion is solitary and shows expansion but is traversed by many strand like bands of bone which give an appearance of trabeculation. It occurs at the ends of the long bones. The enchondroma is a benign tumor of cartilage, is central in location and produces true expansion. The lesions are usually multiple and occur in the small bones of the fingers and toes.

affected bone as the result of stress may result in marked deformations because of loss of the rigidity of the normal bony structure.

Fibrous dysplasia frequently affects the bones of the face and skull. Involvement of the jaws and facial bones results in the production of excessive amounts of dense bone. The antrum may be obliterated. Lesions localized in the orbital region may cause unilateral exophthalmus. In the older literature these changes were termed leontiasis ossea, ossifying fibroma, and fibro-osteoma, but are now recognized as manifestations of fibrous dysplasia. The condition is easily confused with meningioma. Meningioma is rare in children, while fibrous dysplasia usually originates during childhood and progresses slowly over a period of many years. Cherubism is a form of fibrous dysplasia which affects the jaws in the tooth-bearing areas. In many instances no other bones of the body have been involved. While it was formerly believed that the changes were related to the growth and development of the teeth, it now appears that the process is in reality unrelated to the dental structures. Cases have been reported in four brothers and a sister and in one series there was a continuous line of five generations all showing the same abnormality.

Fibrous dysplasia of the skull is usually manifested by irregular thickening of the outer table. Superficial lesions are usually associated with cystlike areas which are ovoid or spherical and may become large in size. Involvement of the base of the skull is not accompanied by expansion, the density of the cortical bone in this region resulting in extensive sclerosis. This accounts for the changes associated with leontiasis ossea. The roentgen manifestations in the skull are not uniform. The cystlike translucent areas typically found elsewhere in the body are uncommon. The lesions are usually associated with widening of the diploe and thinning of the tables. In many instances the skull and face are the sites of extensive new bone formation which causes an increase in the thickness and density of the involved areas. The localization of the skull and facial lesions may be unilateral and restricted to the side of the skeletal lesions when the latter involve only one side of the body. The calvaria, the roofs of the orbits, the wings of the sphenoid bones, the anterior and posterior clinoids, the malar bones, the paranasal sinuses, and the mandible may be affected. Schlumberger, in his excellent monograph on the subject, reports one case each in the parietal bone and occipital bone. A patient seen in the clinic with which I am associated presented a lesion in the posterior portion of the parietal bone. The condition produced a rounded or ovoid area of increased radiance with smooth, irregular borders which were not eburnated. The outer table was more extensively thinned and expanded than the inner. This was best demonstrated in tangential projections and could not be visualized in conventional roentgenograms. Spotty calcification occurred within the radiant area, giving a trabeculated appearance.

Pathology. The lesions are fusiform or spherical and vary in size from a few millimeters to several centimeters in diameter. The surface is usually thin but the cortical bone is intact. The areas are called "cystic" because of the absence of bone and resulting increased radiance roentgenographically, but are not true cysts. The spongy bone is replaced by fibrous tissue and the marrow cavity is filled with a grayish white material of the consistency of cartilage through which are scattered trabeculae of poorly formed primitive bone. The connective tissue is the primary component of the bone lesion, is fairly well vascularized and con-

be precocious puberty in association with the polyostotic lesions It is believed that monostotic fibrous dysplasia and polyostotic fibrous dysplasia are not different stages of the same disease entity but rather are secondary to the action of widely different etiologic factors Proliferation of connective tissue is a basic, nonspecific response of the body to injury and is usually an integral part of the repair process When it occurs in the medullary cavity of bone it is termed osteitis fibrosa It is found in chronic osteomyelitis, osteomalacia rickets prolonged acidosis and the vicinity of primary and metastatic bone tumors It is a characteristic feature in von Recklinghausen's disease (generalized osteitis fibrosa cystica) Paget's disease (osteitis deformans) and systemic diseases in which there is a connective tissue replacement of the marrow cavities and adjacent cancellous bone In von Recklinghausen s disease the fibrosis is secondary to an increase in the rate of bone resorption following enhanced activity of the osteoclasts and decalcification of the bone trabecula due to hyperparathyroidism There is an increase in serum calcium and phosphatase, a decrease in inorganic phosphorus and excessive excretion of calcium and phosphorus in the urine In Paget s disease, osteoclastic activity is less intense and new bone formation is accelerated, the irregular partially calcified trabeculae do not form symmetrical Haversian systems being united in haphazard fashion to form a mosaic pattern The serum calcium and phosphorus levels are normal the serum phosphatase is increased

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In dyschondroplasia or Ollier s disease there are irregularly distributed areas of rarefaction in the diaphyses with unilateral expansion of the metaphyses and thinning of the cortex There is pressure absorption but no periosteal reaction or other indication of new bone formation Differential diagnosis is aided by the fact that in Ollier s disease the condition originated in the cancellous structure in the region of the metaphysis while the lesions in fibrous dysplasia have developed in the cortical portions of the shafts of the long bones In Ollier s disease the roentgenogram reveals longitudinal bony strands of preserved bone trabeculae radiating in fan like fashion at the ends of the involved bones interspersed with rarefied areas caused by cartilaginous deposits There are small heavily calcified globular bodies between these mottled areas The condition arises in early childhood during the period of bone growth As the bone develops the inhibition of bone growth in the affected portion of the diaphyses and the continuous normal bone development in the unaffected areas result in marked deformity and shortening of the extremity With continued bone growth the cartilaginous rests may be displaced farther from the *metaphyseal region into the shaft* *The cortical bone* is expanded and thinned and may even be completely destroyed by the mass of protruding immature irregularly arranged cartilage The periosteum which covers some areas of cartilage appears normal and presents no manifestations of bone deposition Eosinophilic granuloma produces an area of *radiance* without increased density in the lesion itself or at its margins Differentiation from fibrous dysplasia may be difficult or impossible Neurofibromatosis may be differentiated by the fact that the lesion in the bone appears to develop by erosion of the fibrous nodule from the periosteal surface into the cortex The associated soft tissue tumors in the skin and subcutaneous structures the generalized distribution of the lesions and the involvement of the epiphyseal cartilage in neurofibromatosis aid in differential diagnosis Other less common lesions which must be considered in differential diagnosis include angioma epithelial inclusion cyst adamantinoma chondroblastoma tuberous sclerosis cystic tuberculosis gumma epidermoid osteoid osteoma lymphoblastoma solitary myeloma pyogenic lesions and fungus infection of bone Biopsy is often essential *for final diagnosis and in some cases doubt remains even after* this procedure

Relationship to Certain Other Similar Conditions It has been stated that fibrous dysplasia of bone is similar to the neurofibromatosis of von Recklinghausen The variability of the connective tissue pattern and the absence of nerve fibers with Bodian s stain are against this theory Lichtenstein suggested that the lesions of the maxilla and mandible which have been described as ossifying fibromas are variants of fibrous dysplasia and present similar histologic pictures It therefore seems that ossifying fibroma and fibrous dysplasia are not separate disease entities although at first glance they appear quite dissimilar Similarly it appears that non osteogenic fibroma of bone which is usually seen in the long pipe stem bones may also be a variant of fibrous dysplasia Polyostotic fibrous dysplasia is characterized by *congenital anomalies* of the bones *and other* organs and excessive cutaneous pigmentation In young girls there may

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ADDITIONAL READING

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OSTEITIS DEFORMANS—PAGET'S DISEASE

Paget's original article appeared in 1877. It was so comprehensive and detailed that relatively little has since been added to his classical description of the clinical aspects of the disease. The roentgen manifestations, certain minor clinical observations and the contributions of biochemistry comprise practically the only additions to our knowledge of the condition since Paget's original paper. Prior to the discovery of the roentgen ray the condition was identified solely on the basis of the deforming features of the disease. The fully developed case can be recognized at a glance. The early stages are impossible to diagnose clinically because similar changes may occur in an old fracture, syphilis, rickets, osteomyelitis and other diseases. Except possibly in the very earliest stages, the roentgen examination establishes the diagnosis of Paget's disease with definiteness. Osteitis deformans is limited to the skeleton. The condition is slowly progressive and has a tendency to eventually involve the entire skeleton. The process may remain stationary for long periods of time and for this reason many authors have described a monostotic form. The disease is not systematic in its distribution and appears to involve any bone in the body with the possible exception of the phalanges of the feet and the hands.

Etiology. The incidence cannot be established with certainty because of the insidious onset. It has been estimated that approximately 3 per cent of adults over forty have Paget's disease. In the leading clinics and hospitals in the United States there appears to be one case of Paget's disease for approximately every 10,000 admissions. The ratio between white and colored is about that of the population in general. While it has been reported in the young, the juvenile cases can hardly be considered authentic. The youngest patient definitely proven to have the disease is about twenty-seven years of age. The largest number of cases occur in the seventh decade. Osteitis deformans has been found in all races and in all parts of the world. The disorder is not neoplastic. Syphilis can be eliminated as the cause. Geography, climate, race, heredity, diet and endocrine influences do not seem to play a part. Some writers have believed that it is familial. The disease is not known in domestic animals but has been described in monkeys and possibly in rabbits. Paget's disease has never been produced experimentally. The irregular distribution eliminates it from conditions or diseases which involve the skeleton generally and systematically. The early changes in the marrow are considered by some to be an indication of an infection or an irritation. The frequent development of sarcoma in Paget's disease suggests an increased proliferative activity of the medullary connective tissues of the bones which when exaggerated markedly results in malignant growth. The classification of Paget's disease as a hyperplastic osteitis appears to be justifiable in the light of our present knowledge. Many authorities consider that the disease is the result of a metabolic disturbance.

Symptoms. The deformity is unlike that in any other bone dystrophy. There is occasionally bone pain but this is usually not as marked as in primary or metastatic neoplasms and infections of the bone. Only a portion of a bone or bones may be affected. Depending upon the region involved, various complications may ensue. The optic nerve may be compressed with resultant blindness. Involvement of the temporal bone



FIG 213 Incomplete Fracture of the Shaft of the Femur in Paget s Disease The femur shows the characteristic changes of Paget s disease The fracture is incomplete and extends horizontally



FIG 214 Paget s Disease The bones of the spine and pelvis present increased density with marked coarsening of the trabeculations There is an actual increase in the size of the bones

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Fig 216 Osteoporosis Circumscripta Paget's Disease There is osteoporosis circumscripta involving the frontal and parietal bones The osteoporotic area is sharply delimited its margins being serpentine in character The tables of the skull are increased in density and thickness

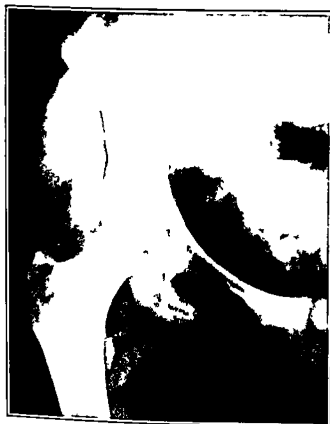


Fig 217

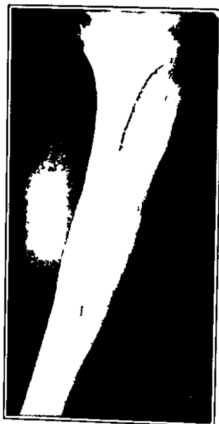


Fig 218

Fig 217 Paget's Disease with Sarcoma There is an extensive area of destruction involving the medial aspect of the right ilium The bones of the pelvis show the characteristic changes of Paget's disease The diagnosis of fibrosarcoma was proven by biopsy

Fig 218 Fibrosarcoma of the Tibia associated with Paget's Disease The shaft of the tibia is increased in density and thickness There are multiple areas of rarefaction in the upper and middle thirds of the shaft of the tibia indicating localized bone destruction Biopsy revealed sarcoma in Paget's disease

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may produce symptoms of pressure on the eighth nerve Trigeminal neuralgia is not infrequently present and may necessitate removal of the gasserian ganglion Practically any of the cranial nerves may be affected by changes in the base of the skull Enlargement of the maxilla and hard palate may interfere with speech, mastication, and deglutition The voice may be altered The spinal cord may be compressed The spinal nerves are seldom affected Although insidious in onset and slowly progressive Paget s disease usually does not shorten life The bones are thickened and deformed and the patient becomes short and squat with curved back and long arms resulting in an ape like appearance The legs are bowed The calvaria is thickened, a characteristic feature being that the patient has to use progressively larger and larger hats There is no fever No diagnostic changes occur in the urine or feces Since the disease occurs



Fig 215 Paget s Disease There is a very marked thickening of the bones with extensive mottling and irregular areas of increased density Both the inner and outer tables are increased greatly in thickness and density There are multiple striations in the bones The pituitary fossa is small and irregular The patient was a woman forty seven years of age She had noted an increase in the size of her head and complained of headaches

principally in the second half of life it frequently is associated with disorders of that period particularly cardiac and arterial lesions

The involved bones increase in both volume and length The deformity is always opposite the greatest mass of muscle and fascia Where one of a pair of bones is involved as in the forearm and leg the lengthening of one bone may deform its companion The skin and subcutaneous tissues accommodate themselves to the lengthened and altered bone In many instances the bone which is elongated undergoes bending In the upper limbs function is not impaired appreciably In the early stages the bone becomes malleable Because of this it has been postulated that the deformities are produced by weight bearing particularly in the lower extremities and the skull This would not account for deformations of the upper limbs which can better be explained by lengthening of the bone without corresponding increase in the length of the muscles and the

when normal growth or repair is prevented subsequent to inadequate supplies of calcium and phosphorus as in rickets and hyperparathyroidism. The normal values in the serum of adults range from 1.5 to 5.0/100 cc Bodansky units and 3.7 to 13.0 cc King Armstrong units. In children the values are from the upper adult normal to three times as much, the levels being dependent on the rate of growth. (2) Acid phosphatase. The acid phosphatase appears to be formed by the prostatic epithelium. It is normally present in large amounts in the prostate gland whether the gland be normal, hypertrophied, or cancerous. Amounts in other tissues are negligible. The enzyme does not enter the circulation from either a normal or cancerous gland unless, in the latter case, the tumor has metastasized to bone or soft tissues. In Paget's disease elevated levels of acid phosphatase are usually but not always demonstrable. The normal values in the serum of adults are approximately 0.2 to 1.0 cc Bodansky units and 0.6 to 3.0 cc King Armstrong units.

B. *Calcium.* The total calcium in the serum of adults is 9.5 to 11.0 mg./100 cc while in children it is 11.0 to 12.5 mg./100 cc. There is no change in serum calcium level in Paget's disease.

C. *Phosphorus.* The serum inorganic phosphorus in adults is 2.5 to 4.5 mg./100 cc while in children the usual level is 4.0 to 6.5 mg./100 cc. The inorganic phosphorus level in the blood serum is within normal range in Paget's disease.

	Acid Phosphatase	*Alkaline Phosphatase*	*Serum Calcium*	*Serum Phosphorus*
Paget's disease	Normal	Increased 20–40 BU common, 200 BU occasionally	Normal	Normal or slightly elevated
Osteoblastic metastases from prostate	Increased (in [illegible]0%)	Increased 20–30 BU commonly, 100 plus BU occasionally	Normal or slightly down	Normal

Complications. The *common complications are fracture and the development of sarcoma.* Fracture is usually produced by slight violence. If truly spontaneous, coexisting disease should be sought for, particularly sarcoma. The fracture of the long bones is generally transverse. Healing of Paget's bone after fractures or surgical operation is rapid, in some instances the healing being more rapid than the normal. The coexistence of sarcoma and Paget's disease is not unusual. In persons over fifty years of age, the presence of osteogenic sarcoma is presumptive evidence of the concurrence of Paget's disease. Sarcoma of the skull is relatively rare and in a series of 208 cases studied by Kirshbaum, sarcoma developed in 3.8 per cent. Geschickter and Copeland state that in Paget's disease sarcomatous change takes place in the affected bones in 5 to 7 per cent of the cases. The occurrence of *multicentric foci of sarcoma* in bones involved by Paget's disease has been reported frequently. Cerebral extension is rare. The sarcoma in Paget's disease is usually of the fibrosarcoma type. The tumor may also be of the periosteal or osteoblastic variety. The association of two uncommon conditions such as osteogenic sarcoma and Paget's disease suggests that there is more than a *coincidental* relationship between the lesions. It is generally appreciated

fascia, rather than by weight bearing In the later stages, the bone is hard and thick The order of frequency of involvement appears to be the lower extremities, the pelvic girdle the skull, vertebrae, ribs and sternum The upper extremity, the pectoral girdle, and the most distal bones of the upper and lower limbs are least frequently involved There is increased warmth over the involved bones and this comprises a symptom of the disease

Laboratory Findings The serum calcium and phosphorus are usually normal although there is a tendency for the serum phosphorus to be slightly elevated The serum phosphatase level is higher per unit of bone disease in Paget s disease than in any other condition With active, widespread

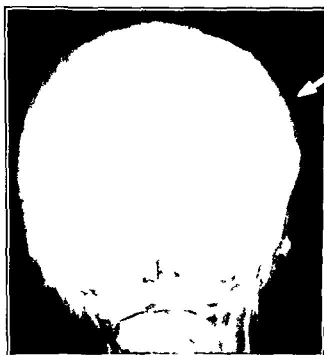


Fig 219 Paget s Disease with Fibrosarcoma The typical changes of advanced Pagets disease are well illustrated with marked thickening increased density of the bones and mottling In the left frontoparietal region there is a rounded area of increased radiance with extensive destruction of the tables (arrow) The patient was a male eighty seven years old who had Paget s disease for many years A swelling the size of an egg had been present in the left anterior aspect of the head for over six months it was soft painless and had been enlarging gradually The patient entered the hospital in coma and death ensued shortly after admission Postmortem studies showed fibrosarcoma arising in Paget s disease

disease exhibiting rapid formation of new bone and increased osteoblastic activity the serum phosphatase may be markedly elevated to 200 Bodansky units and is often about 100 Bodansky units In the early monostotic phase of the disease the elevation may be only slight The elevated phosphatase in the presence of osteoporosis circumscripta associated with Paget s disease is due to the presence of associated foci of sclerosis and not to the osteoporosis circumscripta itself

Blood Chemistry A *Phosphatase* (1) Alkaline phosphatase This enzyme is apparently formed by osteoblasts It is found in increased quantities in the serum whenever bone is being formed This may be normal bone as in fractures or abnormal as in Paget s disease osteogenic sarcoma, or osteoblastic metastases It is elevated in growing bones or

Roentgen Manifestations While Paget's disease is often widespread and affects many bones the skull is one of the sites of predilection and frequently shows earlier and more characteristic changes than other regions of the body In early stages there may be a typical change in the skull, known as osteoporosis circumscripta in which an area of the skull is found to be radiolucent This is an osteolytic or rarefying form of Paget's disease a precursor or atypical form of the disease which usually progresses into typical osteitis deformans The osteoporotic area is sharply delineated involves the outer table first with extension to the inner table later and is usually single although multiple foci may occur An important feature of osteoporosis circumscripta is a narrow band of slightly increased radiance at the peripheral margins of the diffuse rarefaction and represents the advancing edge of the process The borders of the involved portion of the skull show no sclerosis and may be smooth or slightly irregular This is important in the differentiation of intra diploic epidermoid of the skull which presents edges that are scalloped and definitely increased in density owing to expansion and heaping up of the bone There is no evidence of bone regeneration in this phase of the disease which may persist and increase slowly for periods as long as eight years

In the advanced stages, there is marked thickening of the bone with multiple irregular circular areas of increased density and extensive mottling The outer table and diploe are affected first and most markedly The appearance is usually spoken of as 'cotton woolly' or 'cotton tuft in character There is striated appearance due to exaggeration of the trabeculae The base becomes kyphotic and bulges into the cranial cavity The pituitary fossa is small and irregular A significant characteristic of the skull in Paget's disease is that while the outer table is porotic the inner table is sclerotic In many descriptions of the disease the inner table is described as being normal which is actually not the case as the entire thickened calvaria is involved It is important not to confuse the rarefaction of osteoporosis circumscripta with the rarefied areas due to bone destruction in tumors and other lesions The changes usually begin in the outer table with multiple irregular calcific depositions giving the appearance of dense bony islands When viewed tangentially these appear as irregular excrescences and indentations Uneven thickening of the outer table progresses with blending of the outlines of the inner and outer tables Eventually the inner table becomes hazy and indis tinct In the late stages the sutures may be obliterated and the thickness of the calvaria increased several times the normal

The evaluation of the roentgenogram of the skull is important *since* many bizarre, unexplained symptoms and signs referable to varying degrees of cord compression may be clarified by the findings of basilar impression Poppell and his associates carried out a series of observations of the skull in patients with Paget's disease and found a statistically significant trend toward basilar impression 36 per cent of a series of 75 cases showing this anomaly The incidence of platybasia in Paget's disease of the skull on the contrary was not significant *It is important* to have a record of Chamberlain's line and its relation to the odontoid process in all cases of Paget's disease and other malacic or softening con ditions of the bones of the skull as this may give a clue to the presence of cord damage at an early stage It is not unusual in Paget's disease to find excellent visualization of the semicircular canals Newman and Recht-

that Paget's disease must be present for a considerable time prior to the onset of osteogenic sarcoma, the duration usually being about ten to fifteen years The blood alkaline phosphatase levels are not affected by the development of malignant change Biopsy is usually essential for accurate diagnosis Multiple myeloma may also complicate Paget's disease This is of extreme importance as multiple myeloma is more susceptible to therapy than sarcoma From the viewpoint of prognosis and therapy, the establishment of the correct diagnosis is important There is a high incidence of urinary tract stones and of calculi in the salivary glands in patients with Paget s disease

Pathology Paget s disease of the bone or osteitis deformans must be considered an isolated pathologic entity since attempts to classify it either as a tumor or an inflammatory disease are unsuccessful The histologic picture is widely variable and is best described as chaotic The disease does not extend from one bone to another across an articulation On gross examination the affected bone is thickened and presents rough uneven surfaces The compact bone is replaced by spongy bone of varying density and the marrow cavity may be narrowed The difference between the inner and outer tables of the skull frequently disappears and the bone throughout its thickness is entirely made up of spongiosa Areas of sclerosis and porosis are numerous The bending of the long bones and the deformities of the base of the skull have been considered the result of actual molding of the softened bones However there is no evidence that *the bones at any stage of Paget s disease are comprised entirely of osteoid,* and this alone would make such a mechanical deformity possible It appears that in some instances at least, the bending of the long bones is due to a combination of apposition and resorption This view is supported by observation of a curved and elongated tibia in cases in which the fibula is not involved and has remained straight An actual bending can never lead to lengthening of a bone while curving resulting from growth changes necessarily entails elongation

The histologic changes in osteitis deformans involve the bone marrow and the bone tissue The changes in the bone consist of a combination of destruction and repair the two processes being found simultaneously and side by side There is a widely variable ratio of osteoclastic and osteoblastic activity in the affected bone Local differences in this ratio account for the formation of sclerotic and porotic areas in closely adjacent portions of the same bone The resorption proceeds in attacks and these after a variable interval of time are followed by periods of bone formation The alternating sequence of destruction and repair may be repeated in the same regions and leads to the development of a mosaic pattern a characteristic feature of Paget s disease The type of bone which forms in Paget s disase varies widely and is dependent on the speed of its formation In zones of high activity there are large numbers of osteoblasts and bone of an immature coarse fibrillar type is present The bone is thick yet porous There is formation of an inferior type of bone with an irregular coarse matrix The margins of the old and new bone present characteristic curvilinear markings which are typical of the disease There is also periosteal new bone which may reach a thickness of 6 cm or more and is laid down in lamellæ parallel to the periosteum The changes in the marrow are correlated with the changes in the bone tissue and eventuate in fibrosis The fibrosis of the marrow is not reversible

stage of Paget's disease the coarse striated texture of the bone and mottling establish the diagnosis with definiteness. Hyperostosis frontalis interna and diffuse hyperostosis of the calvaria may be confused with Paget's disease. Other conditions which must be considered in the differential diagnosis comprise osteoblastic metastases from carcinoma of the prostate and breast, hyperparathyroidism, carcinoma of the bladder, syphilis, vertebral angiomas, caisson disease and osteopoikilosis.

Paget's Disease	*Osteoblastic Prostatic Metastases*
Usual age at onset 55	65
Incidence [illegible] 0.1% [illegible] 3% (p. [illegible])	10-20% (p. [illegible])
X-ray: Width and length of affected bone increased	No change
Bowing and other deformities	None
Infractions and pathologic fractures	None
Associated (advancing) osteoporosis, mottled and irregular	No associated osteoporosis except that of advanced stage
Trabecular pattern coarse but retained	Pattern obliterated
Often involves tibia, radius and ulna also	Rare below knees and elbows
Cortex blurred, streaked and spongy	Cortex dense and well defined
Pseudo cyst formation	None
Osteoporosis circumscripta	No similar finding
Platybasia	Not associated
Chemistry	
Alkaline phosphatase elevated	Alkaline phosphatase elevated
Acid phosphatase normal	Acid phosphatase elevated

Paget's disease and hyperparathyroidism have many similarities and may occur simultaneously in the same patient. The differentiation of these conditions may be aided by the following tabulation.

PAGET'S DISEASE

A. *Clinical Aspects*

(1) Occurs in older people
(2) More common in males
(3) High incidence of sarcoma
(4) Spontaneous fracture is rare in early phase
(5) Renal stones are the exception
(6) Cortical thickening and bowing
(7) No change in serum calcium or inorganic phosphorus
(8) Usually no increase in excretion of calcium and phosphorus; if excretion is increased it occurs in both the urine and feces

B. *Roentgen Aspects*

(1) Increased coarseness and irregularity of trabeculae in enlarged hyperostotic bones
(2) Greatest severity in weight and stress bearing bones. The long bones are bowed
(3) No brown tumors or cysts as a rule

schaffen emphasized that the presence of osteoporosis of the temporal bone in association with marked osteosclerosis of the osseous labyrinths of the inner ears accounts for the sharp delineation of the semicircular canals Impairment of the inner ear occurs in patients with Paget s disease in whom deafness is a complication and the damage to the inner ear is correlative to the changes in the petrous bones

In the long bones, there is an actual enlargement of the bone with increase in width and density and bowing The original cortical outline is lost and is replaced by one that is increased in thickness and decreased in density with many irregular trabeculæ Bowing takes place toward the side opposite the greatest muscular bulk and there may be greatly increased density in the cortex along the concavity of the curvature The medullary cavity is markedly decreased in size or obliterated There are wide reticulations which usually extend longitudinally or obliquely The sclerotic changes gradually increase and in the late stages the entire bone becomes extremely dense with practically complete absence of normal structure The cancellous bones show marked increase in density and extensive mottling with less marked external thickening than in the long bones Small cysts are apt to occur at the ends of the diaphyses and the region of the epiphyses Pathologic fractures are not uncommon and usually are in the form of thin straight lines extending directly across the bone Healing is rapid and complete as a rule In the pelvis the acetabula appear recessed and deepened with a rachitic type of deformity The spine shows dense wavy irregular trabeculations There may be an actual increase in the size of the affected vertebræ, particularly in the anteroposterior diameter Less commonly there is collapse or narrowing of the vertebræ The intervertebral spaces are usually well preserved Kyphosis is common scoliosis is less commonly seen The changes in the spine and pelvis may be similar in many respects to those in osteoblastic metastatic carcinoma Numerous cases have been recorded in which a single vertebra showed markedly increased density

Fractures are of common occurrence in patients with Paget s disease and frequently occur after slight or trivial injuries Characteristically the fracture is transverse in type and is associated with displacement and separation of the fragments *Incomplete linear fractures* similar to the so called Looser s line are common These may be multiple, two or more being present simultaneously in the same bone or in different bones The long bones showing marked bowing are especially apt to be affected, the incomplete fracture involving the convex portion of the curved bone Healing of fractures in Paget s disease is rapid and complete in practically all cases

The development of sarcoma is usually manifested by the presence of an osteolytic process in the bone The margins of the area of rarefaction may be smooth or irregular and poorly defined Proliferative changes with spicule formations radiating irregularly into the adjacent soft tissues may occur but are less common Repeated roentgen studies at intervals of a few weeks or months show definite progression of the changes A soft tissue mass may be present in the region of the osseous defect particularly in the case of the scalp and less frequently the long bones

Differential Diagnosis In the earliest phase there is rarefaction The rarefied phase can be simulated by loss of overlying soft tissue In the case of the skull the symmetrical thinning of the parietal bone so called hyperplasia of the diploe may produce a similar appearance In the later

THE RETICULOSES

The term reticulosis or reticuloendotheliosis in the recent literature has been limited to a small group of systemic diseases which are linked together by the presence of a predominating histiocytic cell element but in which the etiologic agent cannot be demonstrated. In this group are included Gaucher's disease, Niemann-Pick disease, Schuller-Christian disease, Letterer-Siwe disease and eosinophilic granuloma. Lipoid reticuloendotheliosis is the term used to designate Gaucher's and Niemann-Pick disease. The storage diseases are termed the various types of xanthomatoses. The bones may be involved in these entities which are believed to be a reaction to a specific agent, possibly infectious in origin, although the nature of the agent is unknown. The diseases have in common the presence of a tumorlike or diffuse proliferation of histiocytes. The distribution of the affected nodes may be localized or general. The course may be acute or chronic and either benign or malignant. An important characteristic is that the severity may be correlated in a measure to the age at which the condition occurs. Many stages of these entities may result fatally. There is a marked variation in the character and distribution of the lesions.

There is no universally acceptable classification of the reticuloendotheliosis. This is due to many factors. Tumors of the reticuloendothelial system structurally overlap the endotheliomas and the blastomas of the lymphoid system. An additional complicating factor is that elements of the reticuloendothelial system and the hematopoietic system have the same progenitors. The situation is further complicated by the existence of purely hyperplastic reactive reticuloses which makes it impossible to differentiate between the benign and malignant forms of the disease. Walthard suggested a histopathologic classification based on the pluripotency of the primitive mesenchymal cell since a single mother cell may give rise to markedly complex structures. His classification comprises all elements of the reticular system and includes the connective tissue cells, reticulum cells, endothelial cells and blood cells. He also includes the functional, regressive, heterotopic and inflammatory changes, the generalized reticuloses and the malignant neoplasms of the reticular tissues. The organs most commonly involved are those which under normal conditions contain large amounts of histiocytes or macrophages and comprise the lymph nodes and the lymphatic follicles, the spleen, the thymus, the tonsils, the liver, the skin and the bone marrow. Diffuse accumulations of histiocytes are also found in the connective tissue of the lungs and heart and in some of the endocrine glands. Involvement of the bone marrow and consequently of the skeleton is a common factor in all types of these diseases of the macrophage system.

Niemann-Pick's Disease

Niemann-Pick's disease is a type of lipidosis which occurs in early infancy. It is characterized by the presence of sphingomyelin in the histiocytes. Bone lesions are rare and are not as a rule sufficiently characteristic to permit of the establishment of the diagnosis by roentgen methods alone.

(4) Skull thick, moth eaten, and shows osteoporosis circumscripta
(5) No change in lamina dura

HYPERPARATHYROIDISM

A *Clinical Aspects*
(1) Occurs at all ages, more in females
(2) Sarcoma rare
(3) Fractures common, especially in brittle bones
(4) Nephrolithiasis in 85 per cent
(5) Bone shows thin cortices, cysts and brown tumors characteristically
(6) Hypercalcemia and hyperphosphatemia with resultant gastrointestinal symptoms
(7) Increased excretion of calcium and phosphorus in urine but not in the feces This causes renal calcifications polyuria and polydypsia which may simulate diabetes insipidus
(8) Palpable tumor in neck in 10 per cent
(9) Epulis is common

B *Roentgen Aspects*
(1) Sparse irregular trabeculae in osteoporotic bone
(2) Lesions are generalized
(3) Scattered bone cysts in 40 per cent Brown tumors and pseudo cysts are common
(4) Skull Finely granular ground glass appearance areas of rarefaction and indistinct diploe no thickening of tables
(5) Lamina dura absent a manifestation of generalized decalcification

Therapy No treatment is effective It should be stressed that there is distinct danger in immobilizing the patients for prolonged periods Under these conditions there are no stresses to stimulate bone production while destruction continues unabated The treatment for malignancy superimposed on Paget s disease is uniformly unsuccessful the disease proving fatal in all instances with no proven five year cures All cases of Paget s disease should be observed periodically in order to detect malignant changes as early as possible Marked deformities of the long bones may require surgical correction

Prognosis The prognosis as to life is good The complications such as optic nerve and spinal cord compression are more serious and may alter the prognosis The development of sarcoma is of the gravest prognostic import

THE RETICULOSES

The term reticulosis or reticuloendotheliosis in the recent literature has been limited to a small group of systemic diseases which are linked together by the presence of a predominating histiocytic cell element but in which the etiologic agent cannot be demonstrated. In this group are included Gaucher's disease, Niemann-Pick disease, Schüller-Christian disease, Letterer-Siwe disease and eosinophilic granuloma. Lipoid reticuloendotheliosis is the term used to designate Gaucher's and Niemann-Pick disease. The storage diseases are termed the various types of xanthomatoses. The bones may be involved in these entities which are believed to be a reaction to a specific agent, possibly infectious in origin, although the nature of the agent is unknown. The diseases have in common the presence of a tumorlike or diffuse proliferation of histiocytes. The distribution of the affected nodes may be localized or general. The course may be acute or chronic and either benign or malignant. An important characteristic is that the severity may be correlated in a measure to the age at which the condition occurs. Many stages of these entities may result fatally. There is a marked variation in the character and distribution of the lesions.

There is no universally acceptable classification of the reticuloendothelioses. This is due to many factors. Tumors of the reticuloendothelial system structurally overlap the endotheliomas and the blastomas of the lymphoid system. An additional complicating factor is that elements of the reticuloendothelial system and the hematopoietic system have the same progenitors. The situation is further complicated by the existence of purely hyperplastic reactive reticuloses which makes it impossible to differentiate between the benign and malignant forms of the disease. Walthard suggested a histopathologic classification based on the pluripotency of the primitive mesenchymal cell since a single mother cell may give rise to markedly complex structures. His classification comprises all elements of the reticular system and includes the connective tissue cells, reticulum cells, endothelial cells and blood cells. He also includes the functional, regressive, heterotopic and inflammatory changes, the generalized reticuloses, and the malignant neoplasms of the reticular tissues. The organs most commonly involved are those which under normal conditions contain large amounts of histiocytes or macrophages and comprise the lymph nodes and the lymphatic follicles, the spleen, the thymus, the tonsils, the liver, the skin and the bone marrow. Diffuse accumulations of histiocytes are also found in the connective tissue of the lungs and heart and in some of the endocrine glands. Involvement of the bone marrow and consequently of the skeleton is a common factor in all types of these diseases of the macrophage system.

Niemann-Pick's Disease

Niemann-Pick's disease is a type of lipidosis which occurs in early infancy. It is characterized by the presence of sphingomyelin in the histiocytes. Bone lesions are rare and are not as a rule sufficiently characteristic to permit of the establishment of the diagnosis by roentgen methods alone.

Letterer-Siwe's Disease

Letterer Siwe's disease affects infants and children below the age of two years. It runs an acute or subacute course, usually terminating fatally in weeks or months. In rare instances the duration is one or two years. The condition is characterized by enlargement of the liver and lymph nodes, purpura, secondary anemia, a normal or low white count, and the appearance of destructive lesions in the bones, the skull being most commonly affected.

Gaucher's Disease

Gaucher's disease occurs most commonly in young Jewish females. It is a hereditary or familial disturbance and is characterized by bony xanthomas and the deposition of lipoprotein in portions of the reticulo endothelial system. There is a low platelet count, tendency to hemorrhage, and enlargement of the spleen. Although similar to Schuller-Christian's and Niemann Pick's disease, it is differentiated by the presence of the so called Gaucher cells, reticuloendothelial macrophages, cells containing kerasin rather than cholesterol or lecithin. The condition was first described by Gaucher in 1882. In 1904 Brill and his associates found the Gaucher cells in the bone marrow in autopsy material. On microscopic examination the cells appear as minimal collections, as nodules, or as a diffuse infiltration in the bone marrow. The extent of the infiltration of the abnormal cells determines the extent of the related lesions which are demonstrable on x ray examination. In many instances roentgen studies establish the diagnosis with a high degree of accuracy.

The bone lesions of Gaucher's disease are due to infiltration and replacement of the bony trabeculæ by kerasin bearing reticulum cells, the so called Gaucher's cells. Gaucher cells have been found only in the spleen, the liver, the bone marrow and the lymph nodes, never in other organs. Clinically the disease is manifested by marked enlargement of the spleen and liver, brownish pigmentation of the exposed parts of the skin, wedge shaped thickening of the conjunctiva near the cornea, a severe hemorrhagic tendency, leukopenia and thrombocytopenia. The diagnosis is established by splenic or bone marrow puncture or by splenectomy. The onset is usually in childhood and adolescence. The condition is chronic and slowly progressive, most of the patients living a normal life for many years. There are periods when there are local manifestations alternating with intervals during which there is almost complete freedom from symptoms. Cases have been reported in which the bone lesions have apparently regressed. The process is particularly apt to regress in areas not subjected to severe stress, as for example the iliac wings. Therapy consists of splenectomy, although removal of the spleen does not arrest the progress of the disease or prevent bone involvement. It relieves the patient of a burdensome spleen and eliminates the tendency to hemorrhage. In several reported cases the changes in the bone have appeared soon after splenectomy. It is presumed that the skeletal changes are manifestations of an aseptic necrosis, the stress of weight bearing and inadequate anatomical reduction and fixation of fragments after pathologic fractures. When changes are present in the lumbar vertebræ there is a slow progression during a period of many years with narrowing of the intervertebral spaces. *In the sacro iliac joints there is mottling and in*creased density in the regions adjacent to the joint with partial or com

plete obliteration of the joint space. The symphysis pubis may be affected and appear partially dislocated.

Roentgen Findings. The skull on roentgen examination shows generalized decrease in density with apparent bone absorption and marked osteoporosis. There are no localized areas of destruction as in Schuller-Christian's disease. In the long bones, particularly the lower end of the femur, there is marked generalized decalcification, thinning and expansion of the cortex, and formation of cystlike areas with partial or complete absence of bone structure. The femur shows the most typical changes and the process may appear limited to the *femora*. *Fusiform expansion of*

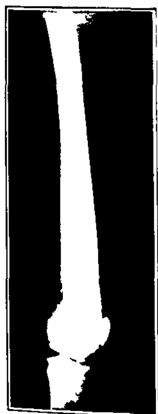


Fig. 220



Fig. 221

Fig. 220. Gaucher's Disease. There is marked rarefaction, expansion and cortical thinning *involving the lower femur. The trabecular pattern is obliterated.*

Fig. 221. Gaucher's Disease with Bone Formation.

the lower ends of the femurs with rarefied areas of varying sizes and scattered patches of osteosclerosis occur. Compression fractures may ensue. The head and neck of the femur and the bodies of the vertebrae may show areas of bone destruction; the hip and intervertebral joint spaces are preserved. The cortex of the long bones is markedly thinned and the medullary canal is widened. Expansion of the femoral diaphysis just above the epiphyseal line may result in a club shaped deformity.

In certain cases of Gaucher's *disease there is bone formation* in the medullary cavity. The new bone is moderate in amount and is similar in appearance to the regenerative ossification which occurs in inflammatory lesions. The changes develop not only in the portions of the bone which show absorptive changes but also in areas which present no roentgen

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twenties, the thirties or later. There is a characteristic triad of symptoms with exophthalmus, diabetes insipidus, and circumscribed defects in the cranial vault. The exophthalmus is the result of extension of the lesions into the orbit. The diabetes insipidus is due to involvement of the hypophysis and the tuber cinereum. In many instances the classical triad of symptoms is incomplete. In others, involvement of various organs results in more conspicuous manifestations. Early degeneration of the hypophysis may lead to dwarfism and hypogonadism. The first manifestation of the disease may be a cystic defect in the jaws. The growth of the nodes may result in loosening and loss of the teeth. Practically any bone in the body may be the site of the histiocytic proliferation. There may also be involvement of other organs, the lymph glands, the spleen, the liver, the



Fig 222. Xanthomatosis. There are multiple large, rounded and irregular areas of increased radiance with punched out borders scattered throughout the skull. There are no sclerotic changes at the borders of the lesions although the uninvolved portions of the skull are dense, indicating osteoblastic changes.

skin and the lung. The disease runs a chronic course, often extending over a period of ten to fifteen years. Fatal termination is usually due to secondary involvement of the hypothalamus or the heart. The patient may suffer from jaundice, malnutrition, anemia and cutaneous xanthomatosis with multiple nodules under the skin, especially in the eyelids and near the outer canthus of the eye. Soreness of the mouth and loosening of the teeth are due to deposits of lipoid in the gums. Cases occur with bone defects alone, other symptoms being absent. The bone defects are due to granulomatous deposits filled with cholesterol and may be of very large size. The typical findings in the bone lesions are the so called foam cells, histiocytes filled with lipoid.

Roentgen Manifestations. The bone defects consist of round, oval or markedly irregular areas of increased radiance which have sharp, clearly-defined margins. The lesions in the flat bones are punched out, present

manifestations of bone destruction The new bone formation occurs only in the later stages of the disease Associated abnormalities comprise changes in the shape and structure of the head of the femur, collapse of the vertebral bodies with gibbus, and pathological fractures In certain instances mottled areas of bone erosion alternate with zones of condensation Windholz and Foster describe two cases with marked new formation of bone The distal portions of the femurs showed diffuse bone formation intermingled with translucent areas and fusiform expansion Bands of ossification were present in the medullary region The bands measured about 5 mm in width were separated from the inner aspects of the cortex by a distance of 1 to 2 mm, and extended the entire length of the shaft of the femur The process of osteogenesis in Gaucher s disease was first described by Pick It appears that ossification in the bone marrow in Gaucher s disease is not an incidental finding, but is the product of reticuloendothelial proliferation developing as direct osseous metaplasia of reticular and collagenous fiber No evidences of bone formation occur as early skeletal manifestations of Gaucher s disease At this stage only the absorptive or destructive phase are present the bone productive phase being a late development The appearance of bone in the spongy structures adjacent to areas of bone erosion may be misinterpreted as evidence of osseous regenerations in the vicinity of an inflammatory lesion This explains the fact that the similarity of the bone changes in Gaucher s disease to those of osteomyelitis and chronic tuberculosis has been stressed by many authors

In children the head of the femur may be affected and show changes similar to those in Legg Perthe s disease Infiltration of the bone by Gaucher s cells produces radiolucent areas in the head and neck of the femur The head of the femur usually shows increased density the changes being those of aseptic necrosis probably due to masses of Gaucher cells in the lumens of the veins and fine capillaries It must be stressed that in some cases of Gaucher s disease the bone changes are the predominant ones

Differential Diagnosis It is necessary to rule out leukemia, cirrhosis of the liver hemolytic jaundice splenic anemia Banti s disease, and thrombosis of the splenic vein The bone lesions may be confused with many diseases the chief of which are multiple myeloma chloroma anemia with sclerosis of the skeleton, osteolytic bone tumors eosinophilic granuloma Schuller-Christian disease and Letterer Siwe disease In Letterer-Siwe's disease the process develops before the age of two years runs an acute or subacute course and is fatal in a few weeks or months The destructive bone lesions affect chiefly the skull In Schuller Christian disease the onset is usually between the ages of five and ten years The lesions occur chiefly in the optic or pituitary fossas and the bones of the skull which accounts for the classical symptoms of exophthalmus diabetes insipidus and localized skull defects There is a chronic course of ten to fifteen or more years and death is usually secondary to involvement of the heart or hypothalamus Eosinophilic granuloma arises in the marrow of one a few or many bones It is benign and may heal spontaneously or after simple removal

Hand-Schuller-Christian's Disease

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twenties the thirties or later. There is a characteristic triad of symptoms with exophthalmus, diabetes insipidus, and circumscribed defects in the cranial vault. The exophthalmus is the result of extension of the lesions into the orbit. The diabetes insipidus is due to involvement of the hypophysis and the tuber cinereum. In many instances the classical triad of symptoms is incomplete. In others involvement of various organs results in more conspicuous manifestations. Early degeneration of the hypophysis may lead to dwarfism and hypogonadism. The first manifestation of the disease may be a cystic defect in the jaws. The growth of the nodes may result in loosening and loss of the teeth. Practically any bone in the body may be the site of the histiocytic proliferation. There may also be involvement of other organs, the lymph glands, the spleen, the liver, the



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There is no mental deterioration. There is a definite correlation between the age of the patient and the prognosis. The younger the patient at the time of onset, the poorer the outlook. There is a tendency to recovery in some cases and death in others [illegible] clear.

Pathogenesis. The clinical manifestations are obscure and in many instances the diagnosis is not established despite the fact that the patient is under observation for long periods of time. The diagnosis can be made by roentgen methods with a high degree of accuracy. Pathologic study of biopsy or necropsy material may be inconclusive. The condition was first described by Hand in 1893. Schuller in 1915 reported a patient with unilateral exophthalmos, bony defects in the calvaria and typical dystrophia adiposogenitalis. He also described a case with exophthalmos, cranial and pelvic bone defects, a healing pathological fracture of the femur and diabetes insipidus. In 1918 Christian reported a patient with a two-year history of progressive disease. Rowland in 1928 correlated two cases of his own with the twelve previously reported of lipoid reticuloendotheliosis and stated that these were all variations of the same clinico-pathological entity. He based his conclusions on the fact that there was lipid deposit in areas rich in reticuloendothelial cells. Pathologists have for many years known of the presence of certain heteromorphic phagocytic cells which lie loosely in the crevices of tissues where they may wander or whence they may enter the circulating blood stream. These have been variously termed wandering cells, clasmatocytes, adventitial cells, macrophages, histiocytes, endothelial leukocytes and polyblasts. Aschoff noted that these cells occurred in large numbers in certain specific somatic locations and assumed that there was a definite reticuloendothelial system with localized collections of phagocytes and therefore termed them reticuloendothelial cells. The largest collections of reticuloendothelial cells occur in the sinuses of the spleen, the bone marrow, the lymph nodes and the lungs. The Kupffer cells of the liver are morphologically and functionally indistinguishable from the free wandering members of this group. It is known that many ingested fats pass through the intestinal walls without change and enter the blood and lymph streams as undissolved foreign material. They are engulfed by phagocytes of the reticuloendothelial system and are found in abundance in the reticuloendothelial cells. Rowland suggests a mechanism for the development of so-called Hand-Schuller-Christian disease. In the presence of an increased amount of undissolved material such as cholesterol lipids in the circulating blood there is injury to the vascular linings and perivascular infiltration with large numbers of the phagocytic reticuloendothelial cells. The involved areas increase rapidly because of the coalescence of adjacent cells and the ingestion of smaller or damaged cells by the larger ones. As a result there is delay in the capillaries with death of more cells and consolidation into the phagocytes. New phagocytes constantly arrive in the area through the blood stream and result in a characteristic microscopic arrangement of smaller newer cells in the center of the lesions near the vessels with larger cells more full of lipoid material at the periphery. The larger cells are displaced from their blood supply and undergo aseptic necrosis to be replaced by fibrous tissue. This explains the presence of diffuse fibrosis which is a prominent feature of this disease. Wallgreen is of the opinion that lipoid deposit in the phagocytic cells is a secondary and local phenomenon and that idiopathic abnormal infiltration of reticuloendothelial cells is a primary lesion of the disease.

smooth or circinate borders, involve the entire thickness of the bone, and may be single or multiple. The absence of sclerosis about the margins is important in differential diagnosis. In advanced skull involvement, the arrangement may resemble the map of a continent and the appearance is referred to as "geographic skull." In the jaws, the areas of radiance may simulate cysts but they occur at the apices of partly formed teeth in which location cysts do not form. After irradiation, areas of bone sclerosis may develop owing to repair. The common sites are the skull, pelvis, femurs, ribs, humerus and scapula. Involvement below the elbow and knee is unusual and vertebral lesions are rare. As the defect enlarges, it destroys the bone along its margins leaving no normal bone in the center of the area of destruction. The defects show no marginal reaction until after treatment. There is no relationship between the size of the lesion and other factors of the disease. Irregular destruction of the bone results in a trabeculated appearance. While any portion of the shaft may be affected, the proximal and distal segments are the most common sites. The epiphysis is usually spared. As the lesion increases in size, there is destruction of the inner surface of the cortex with expansion and pathologic fractures may occur. After therapy, periosteal reaction is not uncommon and the new bone formation may extend into the adjacent portions of the bone. When the disease involves the mandible, the teeth surrounded by the granulomatous process appear to be floating in the mouth. Both the deciduous and permanent teeth may be affected.

Lesions in the thorax associated with pulmonary involvement are characterized by a fine, nodular density scattered throughout the lungs along the distribution of the bronchovascular markings. In a case reported by Chester and Kugel the pulmonary lesions at autopsy were found to consist of small irregular nodules varying in diameter from 0.5 to 2.0 centimeters. In a second case there was diffuse granulomatous infiltration of the lungs. Therapy given to one side resulted in moderate regression of the changes. After healing takes place, the lesions disappear completely or remain as irregular, linear strands of fibrosis. In patients with severe pulmonary involvement with healing the fibrosis seems to be more prominent in the lower portion of the lung fields.

Differential Diagnosis. The principal diseases which must be considered are metastatic carcinoma, multiple myeloma and similar localized lesions of the skeleton. In the case of the skull it is necessary to consider meningioma, epidermoid cyst and osteomyelitis. Metastatic carcinoma and myeloma may be indistinguishable by roentgen methods. Myeloma is rare in children. In osteomyelitis the destructive process leaves areas of uninvolved bone associated with the destruction, which does not occur in the lesions of xanthomatosis. In the long bones, diagnosis is particularly difficult. The condition may resemble Ewing's sarcoma, metastatic tumor, osteomyelitis, chondroma, giant cell tumor and fibrous dysplasia. Pulmonary changes occur only in the presence of associated lesions in the skeleton, although rare exceptions may occur. The most important feature in diagnosis is the appearance of the cranial bones on roentgen examination.

Prognosis. The average duration of the disease is slightly over two years although it may last six or more years. While one lesion is healing others may develop elsewhere in the skeleton. The new site may be near or far removed from the original area. New lesions may develop rapidly during a period of two to six weeks. In general, the prognosis is good

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Fig 223 Eosinophilic Granuloma of Skull There is a localized area of radiance with slightly irregular borders in the temporoparietal region There is no rim of calcification about the edges of the area as occurs in the cholesteatoma or epidermoidoma There are no bony spicules within the lesion The area suggests a metastatic process However the age of the patient is against this possibility

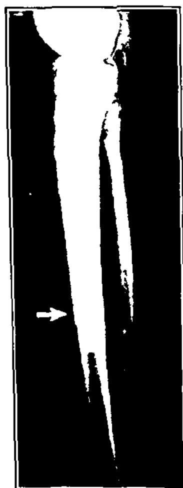


Fig 224 Eosinophilic Granuloma There is a rounded area of radiance in the middle third of the shaft of the tibia The margins of the lesion are irregular and poorly defined There is no sclerosis of the surrounding bone or reaction in the periosteum The roentgen diagnosis of eosinophilic granuloma was confirmed by histopathologic study of the resected specimen

Treatment Early diagnosis is important As soon as the diagnosis has been established, radiation therapy to every organ or region which manifests signs of disease is indicated The disease shows a strong tendency toward spontaneous remissions Irradiation is essential to induce regression Irradiation therapy is best given in small divided doses to the involved areas The dosage to the individual lesion is much less than the cancer dose and adequate therapy can usually be given without producing permanent changes in the skin or other adjacent organs Roentgen therapy is the only satisfactory form of treatment and results in improvement in practically every case After x ray therapy areas of osteosclerosis develop due to repair and osteoblastic reactions Similar changes may also occur in older patients without irradiation During the roentgen treatment new lesions may form in the bone(s) while others are undergoing healing In children the advent of puberty sometimes effects a complete cure

Eosinophilic Granuloma

Eosinophilic granuloma is a destructive bone lesion characterized by large mononuclear cell histiocytes and collections of eosinophils It occurs most frequently in children and young adults and more often in males than in females Trauma virus infection and inflammatory changes have all been considered as etiologic factors While it was formerly believed that the condition was limited to the bones cases with skin lesions have been recorded Pulmonary lesions and diabetes insipidus with involvement of the lymph nodes and the gingiva have been noted In the chest there may be linear, strand like densities and prominence of the markings in the upper lung fields The condition is relatively benign Dickson reports a case of eosinophilic granuloma of bone with characteristic lesions in the acromion process sacrum ischium greater trochanter and vertebrae In association with these lesions there was a diffuse nodular infiltration of both lungs Roentgen therapy to the bones resulted in definite improvement but the lung lesions revealed no response The blood chemistry (cholesterol serum lipid calcium and phosphorus) and bone marrow studies are negative Radiation therapy to the affected bones results in cure The lesions may be single or multiple are most common in the flat bones and have been seen in the skull pelvis vertebrae, ribs and long bones They have been found in all bones except those of the hands There may be no symptoms the process being discovered on routine roentgen study In other cases pain tenderness swelling muscle spasm and atrophy fever weight loss high white count and anorexia may be noted clinically The course is benign Spontaneous recovery is common and the lesion sometimes resolves after several months whether treated or untreated Biopsy for diagnosis usually gives relief of pain Curettage and x ray therapy have resulted in cures

Pathology On gross examination there is a cystic area in the bone usually in the central portion The lesion is composed of soft hemorrhagic or brownish granulation tissue which is streaked with yellow necrotic material Fluid is present within the lesion and may be clear or turbid In the early stages the lesion may be cystic granulomatous and hemorrhagic containing soft yellowish brown material Large, pale mononuclear cells with granular cytoplasm occur in sheets and appear foamy Eosinophils are scattered in clumps or singly in the early stages

Chapter

6

Bone Changes Due to Chemical Substances and Physical Agents

Introduction Metallic salts such as lead phosphorus bismuth and radium may be deposited in bone along with calcium. Prior to fusion of the epiphyses the deposits are most marked on the diaphyseal side of the epiphyseal line and narrow bands of increased density are demonstrable on the roentgenogram. These changes may be present in newborn infants whose mothers have received bismuth injections during pregnancy. The shadows usually disappear within a few weeks or months after the administration or ingestion of the offending agent has ceased. *In adults the foreign material is distributed throughout the bone and casts no shadows on the roentgenogram hence roentgen study is not of value in diagnosis.* Fluorine poisoning occurs in miners working in mines with a high fluorine content in the minerals and also occasionally in persons who drink water containing fluorine. X-rays and radium may produce profound changes in the bones. These manifestations are particularly important in that they may occur without visible alterations in the skin and are frequently insidious in onset.

LEAD POISONING

The clinical picture of lead poisoning has been known for many centuries. In European countries particularly France lead contaminated wine has been the cause of many severe epidemics. Benjamin Franklin refers to cases of lead poisoning from drinking New England rum due to the fact that the rum was distilled in apparatus composed of lead. The chewing of paint from cribs toys furniture, walls and window sills and eating painted plaster and paint flakes are common causes of plumbism. Children of teething age living in old run down houses where paint has been present for many years are particularly apt to show lead poisoning. Putting things in the mouth is normal in the first year of life but is abnormal if continued beyond this period and is referred to as pica or perverted appetite. Many cases of lead poisoning in children are the result of abnormal appetites which have led the patient to ingest dirt of various types and also to the persistence of sucking and chewing things that they can put into the mouth. The incidence of cases of this type decreased markedly after the danger of lead toys was stressed in the medical literature and the use of lead paint on woodwork and children's furniture and toys was abandoned. The public has learned to avoid the use of lead in nipple shields, food wrapping materials cooking utensils and pipes for drinking

and may be absent in the healing stages There may be a few lymphocytes, plasma cells, and polymorphonuclear leukocytes but there is no abscess formation Foam cells occur in the intermediate phases of the disease In the late stages, connective tissue replaces the lesion and new bone formation takes place Letterer Siwe's disease is an acute and more generalized, Hand Schuller Christian's disease a more localized type of the same lesion The relation of eosinophilic granuloma to the other types of histiocytic proliferation is now accepted

Roentgen Findings There is an oval or circular area of bone destruction with little or no bone reaction about its margin The defect may be small or large and while frequently solitary may be multiple In the long bones cortical expansion may result If the cortex is perforated, periosteal bone formation occurs The lesion characteristically is osteolytic in nature and presents a punched out area of radiolucency with sharply defined borders A narrow zone of increased density may be present at the periphery of the lesion In the long bones the granuloma begins centrally and destroys the cortex as it expands There may be expansion of the diaphysis with minimal periosteal reaction Extension through the cortex into the adjacent soft tissues is common and pathologic fracture through the involved area may ensue Sequestration is rarely present The most common sites are the skull, pelvis ribs and long bones although the lesion has been found in every bone except the small bones of the hands and feet The ribs and other flat bones show fusiform expansion at the site of the lesion In the long bones there may be periosteal new bone formation and erosion of the cortex The epiphysis metaphysis and diaphysis are involved with equal frequency In the skull both tables are affected The suture lines do not limit the extension of the lesion

Differential Diagnosis In the differential diagnosis it is necessary to include osteomyelitis tuberculosis, syphilis, bone cyst, fibrous dysplasia and giant cell tumor The multiple form of disease may be confused *with lymphoma osteitis fibrosa cystica and neuroblastoma If the lesion* involves the epiphysis the possibility of giant cell tumor must be excluded In the shaft bone cyst can usually be ruled out by lack of expansion of the cortex Other conditions such as Ewing's tumor, osteomyelitis tuberculosis syphilis fibrous dysplasia and metastatic neoplasm must be carefully considered in the differential diagnosis Multiple defects may closely simulate myeloma lymphoma osteitis fibrosa cystica and Schuller-Christian disease The last is characterized by exophthalmus and diabetes insipidus in association with the skull lesions Biopsy is usually necessary to establish an absolute diagnosis The roentgen changes are frequently typical and if the possibility of this disease is borne in mind, the diagnosis may be made with a reasonable degree of assurance

ticularly in adults in whom there is no roentgen evidence of the disease However, small amounts of lead may be present in the urine and feces without producing toxic effects and the excretion of 0 05 to 0 1 mgm of lead per day in the urine is within normal limits It is important to educate the public in order to prevent this serious disease in children Education and the enforcement of measures regulating the use of lead containing paints will result in a material reduction and eventual elimination of lead poisoning in children

Roentgen Manifestations The roentgen examination is of the greatest value in the diagnosis of plumbism in children as the changes are present in practically every instance and are pathognomonic The characteristic

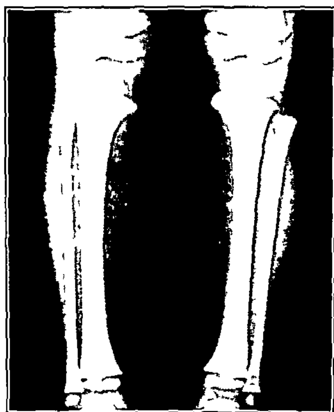


Fig 225 Lead Poisoning There are horizontal bands of increased density at the metaphyseal ends of the long bones The infant was addicted to gnawing the wood of the crib

findings comprise linear bands of increased density extending horizontally across the ends of the diaphyses of the long bones in the juxta epiphyseal regions The opaque bands may also be present at the ends of the flat bones and are due to the deposition of lead in the abnormal and condensed trabeculæ at the growing ends of the long bones and the margins of the flat bones *These manifestations do not occur in adults* After the deposition of lead ceases, a zone of bone of normal density appears on the epiphyseal side of the band and as the bone grows the lead band becomes wider and less defined In the presence of increased intracranial pressure, roentgen examination of the skull may reveal separation of the suture lines and other manifestations of the elevated pressure Roentgenograms of the abdomen may show small specks of radiopaque material distributed irregularly in the gastrointestinal tract due to the presence of metallic lead or lead salts in the stomach and intestines

water Ointments, lotions and dusting powders containing lead applied to the nipples are the source of an occasional case of poisoning in nursing children In the adult poisoning has been caused by hair dyes lead acetate douches, and mouth washes In Japan poisoning in infants was very common as there were no laws prohibiting the manufacture of white lead rouge and toilet and baby powders particularly since Japanese women use large amounts of powder on the face neck shoulders and breasts In many instances the mother absorbs sufficient lead to render the milk toxic to the infant although they themselves present no symptoms In hot dry climates, the use of lead paint on outdoor woodwork has resulted in the inhalation of lead dust and the ingestion of paint This has occurred in children with healthy appetites as well as those with pica

There have been numerous epidemics of lead poisoning in which the source of the lead was traced to the use of storage battery cases as fuel Dealers salvage the lead plates from the batteries and sell or give the cases to friends or needy families The casings are encrusted with lead sulphate and lead peroxide This material burns with an intense but smoky flame Fumes escaping from the stoves and chimneys result in the accumulation of smoke in the living quarters and large numbers of cases of lead poisoning developed in this manner have been reported from Baltimore Philadelphia and other cities Absorption of lead from the respiratory tract is more complete and more rapid than from the alimentary tract or the skin and mucous membranes The liver excretes a large amount of lead absorbed from the intestinal tract In consequence the toxic manifestations after inhalation develop more rapidly and are more severe

Symptoms The symptoms which ensue after the ingestion of lead are very variable in character Loss of appetite vomiting constipation, and cramps are common In children irritability and fretfulness are prominent Severe abdominal pain is frequently present There is pallor and a marked secondary anemia develops Peripheral neuritis and palsy may occur Meningoencephalitis is common Lead lines on the gums and colic are frequent manifestations in adults but are usually absent in patients in the younger age groups Children may present black deposits on the necks of the teeth Severe cases are associated with lethargy projectile vomiting disturbances of vision and alterations in the pulse and respiratory rate Delirium stupor coma elevation of the blood pressure choked discs and optic atrophy may be present and produce a picture which closely simulates brain tumor Unless the condition is recognized early and treatment instituted promptly permanent damage to the central nervous system may result Many cases terminate fatally

The urine contains traces of albumin and glycosuria is often present The cerebrospinal fluid is abnormal and in the presence of encephalopathy, the pressure is elevated Determinations of the amount of lead in the blood and urine give important confirmatory evidence Anemia, reticulocytosis and an excessive number of normoblasts are important in establishing the diagnosis A punctate basophilia in the blood smear is a valuable clue to the disease This manifestation is not constant but is frequently found during the acute stages and in flare-ups due to an infection The presence of stippling in large numbers of the red cells is an indication of lead intoxication The lead may be identified by spectroscopic examination of the blood and spinal fluid Qualitative determinations of the amounts of lead in the urine and stools may be helpful par

changes persist for many years after cessation of the therapy. Normal bone growth displaces the area of increased density from the epiphyseal end of the bone into the shaft. Large doses of yellow phosphorus produce osteoporosis. Microscopic study of the areas of increased density in the growing bone shows the trabeculae arranged in a longitudinal pattern.

FLUORINE POISONING

Fluorine is of particular interest to the dentist as the so-called mottling of the dental enamel is one of the important manifestations of fluorosis. Interest in fluorine has increased as there is an apparently relative immunity to caries in communities where the drinking water contains fluorine. There are many recorded cases of chronic fluorine intoxication in workers in cryolite factories and in certain districts of India. A disease of cattle and sheep grazing in the neighborhood of factories which use fluorine has been described and there is a disease of sheep in the volcanic areas after the eruption of fluorine-containing gas. In communities where the drinking water contains fluorine, changes in the skeleton have occurred only rarely in the United States but are quite common in certain districts in India. The reason for this appears to be the fact that the highest observed content of fluorides, approximating eight parts per million in drinking water, although injurious to the enamel of the teeth, is not sufficient to produce demonstrable alterations in the skeleton. However, these amounts of fluorine may cause osseous changes when they occur in combination with dietary deficiencies, particularly vitamin C or vitamin D, or in the presence of renal damage. The people of the districts in India in which bone lesions have been described doubtless have an insufficient diet.

The changes in spontaneous and experimental fluorosis are apparently contradictory. The bones of human patients show sclerosis. While the experimental studies have in some instances revealed osteosclerosis, there have been other experiments in which there have developed osteoporosis and osteomalacia. These apparent contradictions are believed to be due to differences in dosage and age. Small doses cause sclerotic changes particularly in adults, while large doses produce osteomalacia especially in the young. The apparent contradiction in the results may be due to the presence of calcification in the connective tissue adjacent to the bone. In man the tissues which are affected are the endosteum, the periosteum, the blood vessels and the ligaments. The fact that calcification of the periosteal connective tissue is one of the factors which leads to an erroneous diagnosis of osteosclerosis has been proven by the fact that the bones have a chalky white surface. By roentgen methods it is not possible to differentiate between actual osteosclerosis and the apparent increase in calcium content of the bone due to calcification of the adjacent soft tissues. The same error results if the diagnosis of osteosclerosis is based on observations with reference to the weight of the bone. A diagnosis of osteomalacia appears very doubtful and the most that can be stated is that there is retarded calcification of rapidly produced osteoid tissue.

Histologic studies have been made on animals by Weinman and Sicher. In all animals there is a greatly increased osteoclastic resorption of the premorbid bone. In the shaft of the long bones the compact layer disappears or is broken up into an irregular network of trabeculae. The

In the adult, lead poisoning is of little interest to the roentgenologist because the metal is deposited in amounts sufficient to be demonstrated by roentgen methods only in bone which is undergoing active calcification. Hence, the lead appears only in the metaphyses of children's bones, healing fractures, and bone which is regenerating after radiation therapy of destructive tumors, either primary or metastatic. Since lead-induced poisoning no longer is used in the treatment of cancer, roentgen manifestations of lead poisoning in adults are extremely rare. Roentgen evidence of lead poisoning in children is not infrequent. Prompt recognition of the condition is of the greatest importance since children develop manifestations of lead poisoning after smaller doses and the reactions are more severe. It has been found in epidemics of lead poisoning in which both children and adults have been exposed in the same fashion and were subjected to the same amounts of lead that the children are more uniformly susceptible, especially under the age of four years. There is also a wide variability in the individual response to the same degree of lead poisoning as some persons develop symptoms of toxicity much more readily than do others.

Therapy. Treatment is directed at prevention and immobilizing the lead in the bone. Deleading should not be attempted because of the risk of encephalopathy. Early diagnosis and removal of the cause of the plumbism are of the utmost importance, particularly with infants and young children. The younger the patient, the more severe the manifestations and the more grave the prognosis. Unrecognized and untreated, the disease has a high mortality.

BISMUTH INTOXICATION

Bismuth causes the same changes in the bones of the growing child as lead. Bismuth lines have the identical appearance of lead lines roentgenographically. Bismuth administered to children for the treatment of syphilis may produce bone changes and in mothers treated with bismuth the toxic agent may traverse the placenta and produce bismuth lines in the fetal skeleton. Bismuth changes must be differentiated from syphilitic osteochondritis. The diagnosis of syphilis in the infant should be made only with great caution in cases in which the mother has been treated with bismuth.

PHOSPHORUS INTOXICATION

Phosphorus intoxication may manifest itself in two ways in the bones. Workers in match factories utilizing yellow phosphorus in the past frequently developed persistent osteomyelitis of the mandible. The change in the mandible varied from a mild periostitis to extensive sequestration with involucrum formation. The mandibular involvement represents direct extension of a gingivitis to bone of lowered resistance as a result of the toxic effects of the phosphorus. The substitution of red phosphorus for the yellow variety in match manufacturing has resulted in elimination of the disease. A less common manifestation of intoxication occurs in the bones of young individuals who have not attained full growth. After therapeutic administration of yellow phosphorus in small doses horizontal bands of increased density develop at the ends of the diaphyses. These

tissues Wilkie reported two cases from England in which the patients showed definite osteosclerosis There have been reports by Bishop McMurray Dean and McGarvey and Ernstene of patients showing changes in the bones after exposure to fluorides the excessive intake of fluorides in the drinking water and in persons who had handled sodium fluoride for many years In all of the cases there was evidence of changes in the densities of the bones Largent states that there is a definite correlation between the level of the fluoride concentration in the urine and the presence of changes in the density of the bones He indicates that fluoride concentration of 10 mg per liter is the critical level of fluoride excretion above which an associated increase in the density of the bones may be expected and below which these changes might not be anticipated

Roentgen Manifestations As a rule the changes in the bones are not extensive The roentgen manifestations comprise a slight or moderate increase in the density of the bones The changes may involve the spine the ilia and the sacrum The bones of the thorax particularly the ribs and the long bones may show similar changes In some instances the changes are generalized throughout the skeleton The sacrotuberous the sacrospinous and other ligaments may show extensive calcification The density varies markedly in degree and extent in different bones and in different patients exposed to fluorides Kilborn *et al* report finding arthritis and joint ankylosis in a group of individuals whose drinking water supply showed a fluorine concentration of 6 28 and 5 93 parts per million In one autopsied case a male aged thirty seven years all of the bones were markedly thickened porous rough and brittle There was complete ossification of the intervertebral discs with fusion of the vertebræ the spine being converted into one long rigid bone Roentgen study of the spine revealed increase in density thickening of the trabeculæ and narrowing of the marrow cavities There was questionable rarefaction of the long bones The findings are predominantly those of osteosclerosis although porosis may also occur

In differential diagnosis it is necessary to consider osteoblastic metastases secondary to carcinoma of the prostate In fluorine intoxication the normal bony architecture is preserved and there is uniform distribution of the increased density with calcification of the ligaments Carcinoma of the prostate is associated with disturbance of the bony architecture

destruction of the bone proceeds eccentrically towards the periphery Simultaneously, *there is regenerative and compensatory formation of new* bone which produces a covering of spongy bone which is immature and coarsely fibrillar The formation of osteocytes begins on the periosteal surface of the bone and progresses rapidly, particularly in young animals There are also irregularities in the calcification of the newly formed bone and deposits of mineral salts on the surfaces of the trabeculae The calcification of the osteoid tissue is not homogeneous Rather, there are small and large globular areas of calcification in the intercellular substance In older animals, there is osteosclerosis of the bone but never osteoporosis The bone has a dense spongiosa with thick trabeculations and narrow marrow spaces However, the bone is spongy, does not appear compact and the old shaft of the bone is destroyed by osteoclastic resorption which is active There is progressive destruction of bone with retardation of its calcification In many cases there is extensive damage *to the kidney, particularly if the dosage is high enough to affect the* skeleton The pathologic changes comprise interstitial nephritis with loss of the active secretory tissue It appears that the presence of fluorine brings about rapid resorption of bone but does not hinder the formation of new bone The fluorine changes the intercellular substance of the bone in such a way that there is formation of osteoclasts The rapid resorption of bone appears to commence in the marrow cavities of the long bones indicating that the resorption is an exaggeration of the normal physiologic resorption There is a rapid production of new bone starting at the periosteal surfaces of the shafts of the long bones The new bone radiates from the surface of the old bone and is connected to it apparently as a compensatory strengthening of the bone

The development of osteoporotic changes in young individuals and sclerosis in older persons after a relatively low dosage of fluorine is explained by the differences in the rates of resorption In young animals receiving large doses the resorption of bone is very rapid and the compact bone is destroyed In older individuals the destruction of bone is not so rapid and the formation of compensatory osteophytic bone is proportionately slow Therefore the compact bone is not entirely destroyed but is transformed in part into spongy bone Chronic destruction of bone substance is doubtless responsible for the hypercalcemia and hyperphosphatemia of fluorine intoxication These changes in turn are the basis for the metastatic calcifications of the connective tissues of the bones arteries and ligaments *In some instances there is enlargement of the* parathyroid glands Hypersecretion of the parathyroids secondary to renal damage appears to be common in fluorosis In many respects the *fluorine and the hyperparathyroidism exert a similar action upon the* bone The final picture therefore is a combination of fluorosis and hyperparathyroidism The varying degrees of parathyroid involvement cause *the great variability in skeletal changes under different or in some in*stances under apparently similar circumstances

Roholm reported a series of 10 cases of fluorosis diagnosed during examination *of a group of Danish workmen There was limited mobility* of the spine in some instances and almost complete restriction in 4 of the 10 Another case of practically complete immobilization of the spine in fluorosis was reported by Shortt It appeared that the limitation of mobility of the spine and the changes demonstrated in the vertebral column were subsequent to the deposition of fluorides in the osseous

tissues. Wilkie reported two cases from England in which the patients showed definite osteosclerosis. There have been reports by Bishop, McMurray, Dean and McGarvey, and Ernstene of patients showing changes in the bones after exposure to fluorides, the excessive intake of fluorides in the drinking water, and in persons who had handled sodium fluoride for many years. In all of the cases there was evidence of changes in the densities of the bones. Largent states that there is a definite correlation between the level of the fluoride concentration in the urine and the presence of changes in the density of the bones. He indicates that fluoride concentration of 10 mg. per liter is the critical level of fluoride excretion above which an associated increase in the density of the bones may be expected and below which these changes might not be anticipated.

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In differential diagnosis it is necessary to consider osteoblastic metastases secondary to carcinoma of the prostate. In fluorine intoxication the normal bony architecture is preserved and there is uniform distribution of the increased density with calcification of the ligaments. Carcinoma of the prostate is associated with disturbance of the bony architecture.

THE DELETERIOUS EFFECTS OF RADIATION

The x-ray is a highly destructive agent and extreme caution is essential whenever it is utilized Because the rays are invisible and cause no sensation in their passage into and through the human body, there is frequently a lack of awareness of the fact that harm is being done Even very small amounts of radiation may cause irreparable damage The skin is particularly apt to suffer injury and cancer of the skin is a frequent sequel of repeated small doses in workers with the x ray who have never shown erythema epilation, or other demonstrable effects The damage may not manifest itself until the lapse of an interval of years after the exposure Many of the older x ray workers have atrophy of the skin of the fingers and hands ridging and other alterations of the nails telangiectasis, marked sensitivity of the finger tips to cold, alopecia of the wrists and hands, and other manifestations of carelessness in the use of the x-ray The danger is constantly present and lack of care carries an inevitable penalty Repeated or continuous exposure of large areas of the body may produce leucemia aplastic anemia and other serious blood dyscrasias even though at no time has there been received a dose of sufficient intensity to produce a local reaction

The rapid advances in medical science which have been made in recent years place a great responsibility on the physician This is particularly true in the field of radiation as more powerful machines and innovations in technique create great hazards Engineers and physicists have contributed greatly in the fields of nuclear physics isotopes and allied scientific developments The capabilities limitations and dangers of both the old and the new methods of diagnosis and treatment of disease necessitate a thorough familiarity with the hazards involved and careful judgment in balancing the anticipated benefits against any harm which may result

There is unfortunately an increasing tendency to lose sight of the fact that dangerous and severe biological reactions not infrequently ensue after repeated diagnostic procedures particularly those entailing the use of the fluoroscope The radiologist develops careful techniques and by training habit and instinct learns to prevent harm to his patient and himself The roentgen films and screens in use today are so sensitive and the modern roentgen equipment is so well arranged with regard to protection and the virtual elimination of secondary radiation that the hazards are reduced to a minimum However even with the utmost care and the application of every precaution there still occur occasional instances of erythema ulceration and other serious radiation damage These occur most often from the use of the fluoroscope in the reduction of fractures, the removal of foreign bodies during hip nailing and the insertion of prostheses It is important to note that the incidence of irradiation damage is definitely higher and is apparently increasing in dermatologists surgeons, internists dentists and other members of the nonradiologic groups who utilize the roentgen ray as an adjunct in their practice It is also of the utmost importance to stress the fact that the smaller types of x ray machines have caused the most severe damage There are many reasons for this the chief of which are the less careful shielding of the tube the shorter target screen distance and the longer exposures used in many of the procedures Also a lesser awareness of the potential dangers and lack of care in the application of protective

measures are significant factors. Carelessness in the use of the x-ray and other forms of irradiation has resulted in the untimely deaths of many workers not only in the early years after the discovery of the x-ray but also recently. Equally tragic are the serious damages to the hands, face and other parts of the body of many young surgeons who have been permanently injured and their careers shortened or terminated because of use of the x-ray. In common with other radiologists, cases have come to our attention in surgeons, orthopedists, urologists, internists, dermatologists, and general practitioners of severe, permanent, and crippling injuries by the x-ray.

The dosage required to produce evidences of roentgen damage is dependent on many factors. The kilovoltage, amperage, filtration, length of exposure, size of the field, frequency of the exposure and other variables must be taken into consideration. There is also considerable variation in the degree of susceptibility of various tissues and organs as well as individuals. Blond and light complected persons may show skin reactions with lesser doses than do darker skinned individuals. The international unit of quantity or dose of x-rays or gamma rays is called the "roentgen" and is designated by the symbol "r." "The roentgen shall be the quantity of x- or gamma radiation such that the associated corpuscular emission per 0.001293 gram of air produces, in air, ions carrying 1 esu of quantity of electricity of either sign." Measurements of radiation quantity shall be expressed in roentgens. Measurements of dosage rate shall be expressed in roentgens per minute. With unfiltered radiation generated at voltages less than 90 kvp, dosage of 300 r will produce an erythema with epilation which may be permanent. With a filter of 1 mm. of aluminum, the erythema dose is approximately 400 r. These figures are for areas about 2.5 cm. square. With increase in the size of the field, the back scattering increases the effect and the erythema dose is consequently lessened.

The National Bureau of Standards of the United States Department of Commerce in "Medical X-ray Protection up to Two Million Volts," Handbook 41, establishes the possible dosage rate and maximum permissible x-ray exposure values for various portions of the body. "The maximum total dose to which any part of the body of a person shall be permitted to be exposed continuously or intermittently in a given time shall be 0.300 r per week." On the basis of forty-eight hours per week of uniform exposure, the permissible dosage rates are:

0.00625 r per hour	(6.25 mr per hour)
1.04×10^{-4} r per minute	(0.104 mr per minute)
1.74×10^{-6} r per second	(0.00174 mr per second)

With voltage of 100 kv. and 1 mm. aluminum filtration, the erythema dose is 270 r. The erythema dose, that is, the amount of radiation required to produce a sharp reddening of the skin, is extremely variable. A more accurate standard is the threshold dose, the amount of irradiation required to cause faint bronzing of the skin within two to four weeks after the exposure in the majority of individuals. For unfiltered radiation the threshold dose is 250 r measured in air; with 1 mm. of aluminum filter at 100 kvp. it is 300 r. In the case of large fields, it is necessary to decrease the dose by 10 to 20 per cent or more because of back scattering. Many studies have been made to determine the amount of radiation received by

The x ray is a highly destructive agent and extreme caution is essential whenever it is utilized. Because the rays are invisible and cause no sensation in their passage into and through the human body, there is frequently a lack of awareness of the fact that harm is being done. Even very small amounts of radiation may cause irreparable damage. The skin is particularly apt to suffer injury and cancer of the skin is a frequent sequel of repeated small doses in workers with the x ray who have never shown erythema, epilation or other demonstrable effects. The damage may not manifest itself until the lapse of an interval of years after the exposure. Many of the older x ray workers have atrophy of the skin of the fingers and hands, ridging and other alterations of the nails, telangiectasis, marked sensitivity of the finger-tips to cold, alopecia of the wrists and hands, and other manifestations of carelessness in the use of the x ray. The danger is constantly present and lack of care carries an inevitable penalty. Repeated or continuous exposure of large areas of the body may produce leucemia, aplastic anemia and other serious blood dyscrasias even though at no time has there been received a dose of sufficient intensity to produce a local reaction.

The rapid advances in medical science which have been made in recent years place a great responsibility on the physician. This is particularly true in the field of radiation as more powerful machines and innovations in technique create great hazards. Engineers and physicists have contributed greatly in the fields of nuclear physics, isotopes and allied scientific developments. The capabilities, limitations, and dangers of both the old and the new methods of diagnosis and treatment of disease necessitate a thorough familiarity with the hazards involved and careful judgment in balancing the anticipated benefits against any harm which may result.

There is unfortunately an increasing tendency to lose sight of the fact that dangerous and severe biological reactions not infrequently ensue after repeated diagnostic procedures, particularly those entailing the use of the fluoroscope. The radiologist develops careful techniques and by training, habit and instinct learns to prevent harm to his patient and himself. The roentgen films and screens in use today are so sensitive and the modern roentgen equipment is so well arranged with regard to protection and the virtual elimination of secondary radiation that the hazards are reduced to a minimum. However, even with the utmost care and the application of every precaution, there still occur occasional instances of erythema, ulceration and other serious radiation damage. These occur most often from the use of the fluoroscope in the reduction of fractures, the removal of foreign bodies, during hip nailing and the insertion of prostheses. It is important to note that the incidence of irradiation damage is definitely higher and is apparently increasing in dermatologists, surgeons, internists, dentists and other members of the nonradiologic groups who utilize the roentgen ray as an adjunct in their practice. It is also of the utmost importance to stress the fact that the smaller types of x ray machines have caused the most severe damage. There are many reasons for this, the chief of which are the less careful shielding of the tube, the shorter target screen distance and the longer exposures used in many of the procedures. Also a lesser awareness of the potential dangers and lack of care in the application of protective

measures are significant factors. Carelessness in the use of the x ray and other forms of irradiation has resulted in the untimely deaths of many workers not only in the early years after the discovery of the x ray but also recently. Equally tragic are the serious damages to the hands, face and other parts of the body of many young surgeons who have been permanently injured and their careers shortened or terminated because of use of the x ray. In common with other radiologists, cases have come to our attention in surgeons, orthopedists, urologists, internists, dermatologists and general practitioners of severe, permanent and crippling injuries by the x ray.

The dosage required to produce evidences of roentgen damage is dependent on many factors. The kilovoltage, amperage, filtration, length of exposure, size of the field, frequency of the exposure and other variables must be taken into consideration. There is also considerable variation in the degree of susceptibility of various tissues and organs as well as individuals. Blond and light complected persons may show skin reactions with lesser doses than do darker skinned individuals. The international unit of quantity or dose of x rays or gamma rays is called the roentgen and is designated by the symbol r. The roentgen shall be the quantity of x or gamma radiation such that the associated corpuscular emission per 0.001293 gram of air produces in air ions carrying 1 esu of quantity of electricity of either sign. Measurements of radiation quantity shall be expressed in roentgens. Measurements of dosage rate shall be expressed in roentgens per minute. With unfiltered radiation generated at voltages less than 90 kvp, dosage of 500 r will produce an erythema with epilation which may be permanent. With a filter of 1 mm of aluminum the erythema dose is approximately 400 r. These figures are for areas about 2.5 cm square. With increase in the size of the field the back scattering increases the effect and the erythema dose is consequently lessened.

The National Bureau of Standards of the United States Department of Commerce in Medical X ray Protection up to Two Million Volts, Handbook 41, establishes the possible dosage rate and maximum permissible x ray exposure values for various portions of the body. The maximum total dose to which any part of the body of a person shall be permitted to be exposed continuously or intermittently in a given time shall be 0.300 r per week. On the basis of forty eight hours per week of uniform exposure the permissible dosage rates are

0.00625 r per hour	(6.25 mr per hour)
1.04×10^{-4} r per minute	(0.104 mr per minute)
1.74×10^{-6} r per second	(0.00174 mr per second)

With voltage of 100 kv and 1 mm aluminum filtration the erythema dose is 270 r. The erythema dose, that is, the amount of radiation required to produce a sharp reddening of the skin, is extremely variable. A more accurate standard is the threshold dose, the amount of irradiation required to cause faint bronzing of the skin within two to four weeks after the exposure in the majority of individuals. For unfiltered radiation the threshold dose is 250 r measured in air; with 1 mm of aluminum filter at 100 kvp it is 300 r. In the case of large fields it is necessary to decrease the dose by 10 to 20 per cent or more because of back scattering. Many studies have been made to determine the amount of radiation received by

MAXIMUM PERMISSIBLE X RAY EXPOSURE VALUES

In Milliampere Seconds—at 85 Kvp

FOR ALL PARTS OF THE BODY EXCEPTING THE HEAD

Target—Skin Distance Inches	*Filter (mm Al) External—None Inherent—0 5 Total—0 5*	*Filter (mm Al) External —0 5 Inherent—0 5 Total—1 0*	*Filter (mm Al) External—1 0 Inherent—0 5 Total—1 5*
10	265	510	810
12	380	730	1090
14	520	1000	1500
16	680	1300	1950
18	870	1650	2500
20	1060	2050	3000
22	1280	2450	3640
24	1530	2900	4360
30	2400	4500	6800
36	3460	6500	9800
42	4700	8850	13300
48	6150	11600	17400
	FOR THE HEAD		
10	200	380	610
12	290	550	875
14	390	750	1190
16	510	970	1560
18	650	1240	1980
20	800	1530	2450
22	960	1840	2970
24	1150	2150	3540
30	1790	3360	5540
36	2585	4840	7950
42	3600	6560	10850
48	4600	8550	14100

For distances other than those given in the table, apply inverse square law as the amount of radiation striking a given area varies inversely with the square of the distance from the target of the tube To adjust these tables for kilovoltages other than 85 kvp the following percentage changes may be used

Kvp	*Change in Exposure Values*
100	Reduce by 25%
90	Reduce by 20%
80	Increase by 10%
70	Increase by 35%
60	Increase by 80%

a patient during diagnostic roentgen procedures These are of great importance as untoward reactions occur occasionally after x ray examinations The fact that they occur so infrequently is due in part at least to the rapid loss of radiation effects in the skin It appears that the threshold dose may be repeated after an interval of two to three weeks, about 75 per cent of the dose after one week and 50 per cent in three to four days The recovery rate with low voltage lightly filtered radiation is relatively rapid much more so than after deep roentgen therapy The use of filters is very important and all diagnostic roentgen equipment should have a permanent filter of 1 mm of aluminum permanently incorporated between the tube and the patient This results in a much greater margin of safety The small, shock proof portable type of unit is

particularly dangerous as it is possible to place the tube head in close apposition to the skin thus increasing tremendously the intensity of the radiation to the skin When used with the hand fluoroscope the danger is further increased The surgeon who places his unprotected hand into the direct beam of radiation either during fluoroscopy or the making of a roentgenogram is most apt to suffer harm as the dangers involved in these procedures are particularly great

In evaluating possible damage by the x ray it is important to determine the dosage delivered to the deeper tissues The depth dose at 10 cm below the skin surface for radiation generated at voltages of 100 kvp is approximately 66 per cent of the skin dose measured in air with radiation of which the quality is a half value layer of 0 9 Al and the field size is 250 sq cm To produce an abortion it is necessary to deliver 300 to 360 r to the uterus The temporary castration dose is approximately 210 r delivered to the ovaries These dosages require relatively high voltages and heavy filtration Therefore the effect of the ordinary diagnostic procedures with respect to the production of an abortion or sterility is practically negligible even though a high value of unfiltered or low filtration radiation has been utilized Approximately 20 per cent of an erythema dose delivered to the center of the uterus does not appear to cause abortion although instances have been recorded in which 25 to 30 per cent of a skin dose administered to the uterus has resulted in injury to the fetus despite the fact that the pregnancy progressed to term without apparent deviation from the normal With radiation delivered at 200 kvp and filtration of 0 5 mm copper the skin erythema dose is accepted as approximately 800 r delivered at one sitting hence about 160 r is necessary to cause damage to the fetus Johnson reports a case of agenesis of the cerebral hemispheres in a child who died at the age of six months The mother had received heavily filtered roentgen rays during the early months of pregnancy a dose of about 400 r reaching the uterus Heavy doses of x ray delivered to the pregnant uterus may result in irreparable damage to the fetal central nervous system and optic defects in 50 per cent of the cases In the use of diagnostic radiation, 300 r delivered to the skin results in only 14 r to the uterus and this amount of radiation has never been shown to produce harmful effects

The danger from unfiltered radiation is principally to the skin Modern high milliamperage techniques with increased tube skin distance, intensifying screens and improved roentgen film result in a reduction of the amount of radiation delivered at the skin The depth dosages in diagnostic roentgen procedures are so small as to be negligible with reference to possible effects on the gonads or the pregnant uterus

Irradiation Injuries to the Hand The increasingly high incidence of irradiation dermatitis presents a very difficult therapeutic problem The changes are insidious and may not develop for months or years after exposure Irradiation damage may be divided into 4 groups (1) those in which the exposure to irradiation consisted of many small or minute doses the periods of exposure extending over a period of months or years These comprise the occupational or vocational hazards (2) a massive overexposure usually delivered accidentally These occur commonly after prolonged fluoroscopy in the search for a foreign body diagnostic fluoroscopy the commercial use of the fluoroscope hip nailings and reduction of fractures (3) irradiation in the treatment of lesions of the skin or removal of the hair and (4) treatment of a deep seated condition

MAXIMUM PERMISSIBLE X-RAY EXPOSURE VALUES

In Milliampere Seconds—at 85 Kvp

FOR ALL PARTS OF THE BODY EXCEPTING THE HEAD

Target—Skin Distance Inches	*Filter (mm Al) External—None Inherent—0.5 Total—0.5*	*Filter (mm Al) External—0.5 Inherent—0.5 Total—1.0*	*Filter (mm Al) External—1.0 Inherent—0.5 Total—1.5*
10	265	510	810
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fluoroscopy. Peller and Pick have made a study of the causes of death in 11,481 physicians who died between January, 1947, and December, 1951. In the five year period, the vital statistics issued by the United States Government reveal that almost 9 per cent of all fatal malignancies in physicians are due to the leucemias, compared with only 3.5 per cent in the white male population. This risk is not limited to radiologists and dermatologists, but is also shared by the large group of physicians who routinely make use of the fluoroscope. The increased mortality, approximately two and one half times higher than expected in all age groups, becomes apparent between the ages of twenty five to thirty four years at the period when increased mortality from other forms of malignant disease is also present. Fatal leucemia has occurred in the past about 8 to 9 times more commonly in radiologists and dermatologists than in other physicians. However, during the past ten years the risk involved in the use of roentgen rays by radiologists has increased little if at all; rather it appears to have lessened or remained unchanged. This is in striking contrast to the situation in the case of general practitioners and diagnosticians, as members of these groups who make use of the fluoroscope have shown that they are exposed to a far greater risk.

It is reasonable to expect that in the near future there will be a markedly higher rise in the ratio of leucemia among non-radiologists using the fluoroscope and x-ray machines for diagnosis and therapy. This is without doubt due to lack of proper protection and carelessness in preventing exposure to irradiation. The leaded apron and gloves are not worn at all in many instances, are not used properly in other cases, or are not of sufficient protective power. The lead glass fabric gown may lessen the effects of the rays but in most cases does not afford sufficient protection. Exposure of the upper thorax, head, neck, and other parts of the body may induce neoplastic changes in the hematopoietic centers. The damage results from the accumulation of repeated doses and, depending on the individual's disposition to cancer, results in the development of leucemia. The blood forming organs and tissues appear particularly susceptible, as many other organs and tissues are also exposed to the x-rays and radium and appear to show no excessive incidence of malignant disease. The minimum latent period of leucemia and other malignant tumors induced by the roentgen ray and radium appears to be less than five years.

Irradiation Effects in Radium Miners. In the mine workers of Joachimsthal, there is a long period of latency in the carcinomas of the lung induced by radon and radium, the average being fifteen to seventeen years and the maximum forty years. This is probably due to the fact that a large part of the inhaled noxious agent is exhaled. However, there is a tremendous excess incidence of cancer in the main organ of deposition, the lung. The duration of the latent period is believed to be due to the intensity of exposure, the route taken by the rays or radioactive substance, the speed with which the noxious substances are eliminated, the degree of susceptibility of the individual exposed to cancer, and the ability of the affected tissue to multiply and grow.

EFFECTS OF RADIATION ON BONE

Radiation in dosages sufficient to be therapeutically effective often produces harmful secondary changes which must be prevented wherever

Irradiated skin shows great sensitivity to other irritants such as chemical, mechanical and bacterial agents. It is very sensitive to subsequent irradiation and the effects of sunlight. It is illogical to treat roentgen dermatitis by further irradiation with radon ointments or beta radiation as has been suggested.

The pathologic changes comprise fibrosis, atrophy, telangiectasis, and obliterating endarteritis. Irradiation carcinoma is more frequent after prolonged minimal exposure to soft alpha and beta rays and is usually of the squamous cell variety. Rarely, there may develop basal cell carcinoma. The incidence of malignancy in irradiation dermatitis of the hands is higher than in other parts of the body. The time factor is important and if the dermatitis lasts sufficiently long, carcinoma eventually develops. The reaction of the skin to irradiation may develop in one of two forms. The acute variety develops soon after exposure to large doses of x ray, radium, or atomic irradiation. The chronic form may develop as a sequel to the acute form or as a delayed response to repeated minimal exposures. There are certain areas of the skin, particularly the finger tips, which tend to become thickened and crack. The nails become brittle and irregular and are susceptible to infection. Carcinoma occurs in small ulcers or keratotic plaques. Therapy comprises removal of the affected area. This must be done early and before ulceration and carcinomatous degeneration have ensued. When the lymph nodes are palpably enlarged, radical dissection is indicated.

Dangers of Hip-nailing. During hip nailings, there is great danger of *over-exposure to the x ray. In checking the position of the nail, it is* necessary to utilize the fluoroscope often for considerable periods of time or take not only anteroposterior but also lateral roentgenograms. In the making of the lateral roentgenogram, the surgeon or an assistant holds the cassette in position during the x ray exposure. During a routine hip nailing procedure, two or more roentgenograms are made. The anteroposterior projection should not entail any exposure as the cassette is in a holder under the operating table and the surgeon and attendants can remain in the lead lined booth during the exposure. This is not feasible in the making of the lateral roentgenogram. In a study reported by Rowe during the exposure of five lateral films, it was found that the person holding the film in place was subjected to excessive irradiation. There was delivered to the body 35 mr, at the belt line 28 mr, to the right side of the neck 4 mr, to the left side of the cheek. The hands received 950 mr, to the right ring finger 125 mr, to the left ring finger, and 130 mr, to the left forefinger. The Atomic Energy Commission recommends that 100 mr in any one day or a total of 300 mr in any one week is the maximum acceptable radiation. From the above observations it is evident that the person holding the lateral roentgen film in position is receiving several times the safe dose. It is always essential to use a lead apron and gloves. It is recommended that not more than two films per day or eight films per week should be held by any one person. Various mechanical devices have been developed recently for maintaining the film in position and these always should be utilized. The same person should not assist the roentgenologist repeatedly or constantly. It is essential that avoidance of over exposure to the x ray be stressed constantly as irreparable damage may result *from carelessness in this regard.*

Leucemia in Physicians. Leucemia has become an occupational risk of increasing significance to physicians, particularly those who utilize

fluoroscopy. Peller and Pick have made a study of the causes of death in 11,481 physicians who died between January, 1947, and December, 1951. In the five-year period, the vital statistics issued by the United States Government reveal that almost 9 per cent of all fatal malignancies in physicians are due to the leucemias, compared with only 3.5 per cent in the white male population. This risk is not limited to radiologists and dermatologists but is also shared by the large group of physicians who routinely make use of the fluoroscope. The increased mortality, approximately two and one-half times higher than expected in all age groups, becomes apparent between the ages of twenty-five to thirty-four years at the period when increased mortality from other forms of malignant disease is also present. Fatal leucemia has occurred in the past about 8 to 9 times more commonly in radiologists and dermatologists than in other physicians. However, during the past ten years, the risk involved in the use of roentgen rays by radiologists has increased little if at all; rather it appears to have lessened or remained unchanged. This is in striking contrast to the situation in the case of general practitioners and diagnosticians, as members of these groups who make use of the fluoroscope have shown that they are exposed to a far greater risk.

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Radiation in dosages sufficient to be therapeutically effective often produces harmful secondary changes which must be prevented wherever

possible. The deleterious effects are particularly important in the case of the lungs, the intestines, the bladder and the bones. Formerly it was believed that adult bone was one of the most radioresistant tissues. It is now known that the soft tissue components of bone, particularly the terminal blood vessels of the Haversian and Volkmann's canals are particularly susceptible to radiation and after exposure to x-ray or radium undergo hyperemia with subsequent obliterative endarteritis. Post irradiation vascular changes are important factors in the causation of fractures. Bone does not tolerate radiation well because of its physical and structural characteristics. It absorbs much more energy per unit of volume than other tissues, in some instances the increase in absorption being as much as 30 to 40 per cent. The effect of the irradiation is enhanced by the secondary radiation produced by the high content of calcium and other minerals. In determining the effect on bone, there are several significant factors. The dosage is influenced by the depth of the bone beneath the skin and the quality and quantity of the radiation. The anatomical position of the bone is important as the phalanx or clavicle receives relatively more radiation because of the thinness of the overlying tissues than the neck of the femur. Rays of shorter wave length cause less secondary radiation and are absorbed to a lesser degree than soft rays. Single massive doses produce greater effects than fractionated smaller doses. There is a distinct delay in the reparative reaction of bone tissue after irradiation. The obliterative endarteritis which ensues adds to the delay. The effect may be progressive, vascular occlusion continuing to develop slowly over a long period of time. The damage to the blood supply prevents or delays the normal reparative processes and gradual progression of the changes may produce necrosis months or years after the administration of irradiation. The alterations may cease at any point between the extremes of complete recovery and necrobiosis with death of the tissues. Ewing divided the effects into five stages. (1) The first stage occurs after light irradiation and comprises a decrease in the vitality of the bone cells with a temporary cessation of the growth capacity. This may be followed by full recovery. (2) After heavier irradiation there develops a productive osteitis with thickening of the bone and fuzziness of the periosteum. This does not disappear completely and permanent damage results. (3) Heavier doses cause sclerosis of the bone with fragility. Fractures may occur. The bone shows increased density roentgenographically. (4) Radiation administered to the limit of therapeutic effectiveness may produce aseptic necrosis and sequestration. Absorption of the dead bone may be delayed for years or never take place. (5) The final stage comprises infection subsequent to osteoradionecrosis. This occurs most commonly in the mandible and the bones of the face after therapy for intraoral carcinoma and will be discussed in detail subsequently.

Pathology. The histopathologic picture of radiation osteitis is widely variable. The early changes comprise periosteal thickening, fibrosis and hyalinization with fibrosis of the marrow. As the reaction becomes more advanced there is sclerosis and obliteration of the blood vessels, fat replacement of the marrow and bone sclerosis with the formation of osteoid tissue. In some cases there is no new bone formation. The osteoblasts are more susceptible to radiation than the osteoclasts. The later stages are characterized by fibrosis and loosening of the periosteum and absence of the endosteal osteoblasts. Progression of the changes

results in aseptic necrosis and sequestration There are fragments of necrotic bone surrounded by foreign body reaction and osteoclasts The necrobiosis and sequestration may be fragmentary or massive The sequestra may persist for many years In other types of aseptic necrosis, repair takes place by creeping substitution of fibrous tissue from the periphery with absorption of the dead bone and gradual replacement by new bone This does not occur in necrosis after irradiation because the reparative elements of the dead bone are destroyed over a wide area and the blood supply is markedly reduced

Radiation Injury to the Femur and Pelvic Bones One of the most common bone changes subsequent to irradiation is necrosis and fracture of the neck of the femur The anatomy of the femur and its blood supply are important factors in the production of these effects The vascular supply to the head and neck of the femur is largely from the arteries in



Fig 226 Radiation Osteitis The neck and upper portion of the shaft of the right femur show extensive rarefaction The patient had received intensive roentgen therapy to the pelvis about two years previously

the capsular ligament and the ligamentum teres The limitation of the blood supply in the proximal end of the femur makes it particularly susceptible to aseptic necrosis and fracture An additional important factor is the variation in length and obliquity of the femoral neck at different periods of life In the adult there is an angle of about 130 degrees between the neck and the shaft In consequence of the widening of the pelvis in the female, the neck of the femur forms more nearly a right angle with the shaft and the thickness of the neck is less Baensch in 1927 cited 2 cases of fracture of the neck of the femur after roentgen therapy for carcinoma of the cervix and ovary Subsequent cases have been reported by many observers In a case recorded by Okrainetz and Biller a spontaneous fracture was thought to be due to metastasis and further therapy was administered The patient succumbed to papillary adenocarcinoma of the ovary two years after the occurrence of the fracture On pathological examination there was no evidence of metastases There was marked thickening of the walls of many of the blood vessels hyaline necrosis obliteration of the lumens and no evidence of new bone forma-

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tion It appears that the lateral fields used in the treatment are more important in producing these changes

Roentgen changes may be demonstrable prior to the development of fracture There is increase in the density of the trabeculae accompanied by areas of rarefaction The head and neck of the femur are affected principally although the upper femoral shaft and the acetabulum may also show similar changes The manifestations closely resemble those of the progressive osteochondrosis in Legg-Perthe's disease, post-traumatic aspetic necrosis and caisson disease The similarity of the changes indicates that these lesions may have a common origin Fractures of the hip the bones of the pelvis, and the sacrum may occur after slight trauma or without evidence of trauma The fractures are unusual in appearance and cause only slight symptoms in proportion to the extent of the roentgen changes in the bone In some instances, there is an incomplete fracture with a zone of sclerosis extending across the neck in the region distal to the head There may be separation and displacement of the fragments In some cases the diagnosis can be made only from the history, microscopic studies or the presence of other areas of radionecrosis in the region of the irradiation There may be mottled rarefying and sclerosing osteitis in the pubic bones with fractures of the rami Impacted fractures may occur The recognition of these fractures is difficult Many instances of recovery with little or no disability have been reported In osteoclastic metastases the changes are more extensive, differ in character, and union does not ensue The development of the roentgen changes has been reported as early as five months and as late as sixty two months following the radiation treatment

Radiation Necrosis of the Mandible A serious sequel of irradiation is necrosis and osteomyelitis of the mandible This complication is common in the treatment of intra oral carcinoma and constitutes a limiting factor in the utilization of treatment The radiation often produces necrobiosis of the bone when given in sufficient dosage to destroy the neoplasm Osteoradionecrosis is characterized by painful prolonged course late sequestration of bone and permanent deformities The process is particularly apt to involve the bones of the face and jaws because of the intimate proximity of these structures to the field of treatment The mandible obtains its blood supply from a single nutrient artery the inferior dental artery and from the periosteum It is formed of dense, compact bone and calcium comprises 85 per cent of its mineral content Because of the high calcium content and the proximity to the skin surface the actual intensity of the radiation may be greater in the bone than in the adjacent soft tissues The osseous damage may occur in the presence of intact skin and mucous membrane Infection and trauma accentuate the effects It is advisable as a rule to remove the teeth in the direct field of radiation and in some instances all of the teeth prior to the administration of roentgen therapy The prevention of osteoradionecrosis is difficult Premature surgical intervention may further damage the periosteum and bone and devitalize bone which otherwise might be conserved A period of ten days should ensue between the extraction of teeth and irradiation Once osteoradionecrosis has developed healing is very slow A bony involucrum such as occurs with acute osteomyelitis does not develop and the sequestrum acts as a support around which fibrous union occurs

Radiation Injury to the Ribs Pohle and Frank report a case of radiation injury in which fractures of the ribs appeared four and one half and eleven years respectively after postoperative roentgen therapy for carcinoma of the breast The recognition of the true nature of the lesion is extremely important in differential diagnosis and prognosis The absence of a history of trauma and the asymptomatic nature of the lesion distinguished it from the usual post traumatic fracture It may prove difficult to differentiate the lesion from a metastatic focus Radiation osteitis fracture is characterized by a transverse fracture slow progression failure to heal and eburnation of the margins of the fragments In metastatic disease the defects are wider and more irregular in outline Radiation osteitis of the ribs associated with pathologic fracture develops insidiously and may appear at varying intervals following the radiation Healing takes place very slowly and the lesion may remain ununited for months or years Fractures of the clavicle ribs and other bones have occurred with doses varying from 1500 r to 6000 r The average dose in patients with fractures of the clavicles and ribs is about 2000 r while in the neck of the femur it has been 3000 r Radium or radon in contact with bone is particularly dangerous

Retardation of Bone Growth after Roentgen Radiation It has been shown by many investigators that bone growth in the young can be retarded or arrested by irradiation with x ray or radium The histopathologic changes in the epiphysis following irradiation comprise degeneration with diminution of the number of primary chondrocytes disruption of the orderly arrangement of the cartilage columns and the development of abnormal foci of calcification at a distance from the normal metaphyseal location Measurements of the long bones following roentgen exposure reveal loss of length with decrease in the diameter of the shaft and the thickness of the cortex In a case reported by Frantz, intensive irradiation was delivered to a massive hemangioma After a period of five and one half years there resulted a 29 per cent lag in longitudinal growth of the extremity The circumference of the thigh and calf on the irradiated side was much smaller than normal The diameters of the mid shafts of the long bones exposed to the x ray were greatly decreased The motions of the knee joint were not affected and roentgen studies revealed that the joint space was normal There is no proof that the epiphyses in the portions of the bone not subjected to radiation undergo increased growth, although some observers are of the opinion that there may be a compensatory hypertrophy because of the injury to the irradiated epiphysis

Cancericidal irradiation in infants and children is often followed by permanent alterations or deformities of certain tissues and bodily structures The deleterious effects usually cannot be prevented if sufficient dosage is delivered to the body to arrest or cure the neoplastic process The effect of irradiation in large doses is primarily on the growing portion of the bone Each epiphysis has a specific and characteristic rate of growth which may be altered by the administration of x rays or radium The effect is dependent upon the age of the patient the dose delivered to the epiphyseal center and other factors With dosage less than that necessary to produce necrosis and death of the irradiated tissues there is usually resumption of growth The renewed growth is abnormal in extent and character with alteration of both the rate and the period during which it occurs In three children afflicted with malignant disease, Murphy and

Behrens describe the alterations which developed after irradiation The patients lived for nine to fifteen years after intensive therapy without clinical evidence of active cancer All developed permanent structural and growth changes The epiphysis which shows the greatest rate of growth is affected most markedly even though it may have received a smaller dosage than an epiphysis which has an inherently slower rate of growth Dosages which do not cause complete cessation of growth may result in alteration of the mechanism of bone growth with resultant deformation of the bony architecture and premature closure of the epiphysis Cancericidal irradiation in infants or children induces permanent somatic tissue changes which are manifested by retardation of bone growth deformity of muscular and bony architecture atrophy and scarring of the skin, and fibrosis of the subcutaneous tissues

Radiation Effects on the Spine Scoliosis may be produced by asymmetrical pressure exerted on the bodies of the vertebrae injury to the epiphyses or application of asymmetric tensions or pressures on one side of the growing spine Engle implanted radium seeds in young goats in the tissues adjacent to the spine and caused suppression of growth of the epiphysis As the animals grew the irradiated vertebrae became wedged showed a decrease in height on the side adjacent to the radium needle and developed curvatures the concavities being on the irradiated side Arkin and Simon produced similar asymmetrical suppression of vertebral epiphyseal growth by the use of interstitial radon seeds and external irradiation Implantation of radon in the lower lumbar region caused stunting of the growth of the iliac crest and the transverse process of the vertebra near the seed In order for asymmetry to occur the seed must be placed near one side of the epiphyseal plate and be of adequate strength

Asymmetrical growth arrests have been produced by the use of a beam of roentgen rays with the radiation directed toward one side of the lumbar vertebra through a posterior portal Within seven weeks after administration of a dose of 1000 r the involved lumbar vertebrae showed wedging with decrease in the height of the vertebrae on the affected side Curvature of the spine did not develop until further growth had taken place The curvature is due to direct action of radiation upon the epiphyseal plate The wedging is not the result of pressure from contractural scarring in the concavity of the curve as the intervertebral discs are wider indicating that there is decreased pressure on this side

Arkin and his co workers report a case of scoliosis in a patient whose spine had been heavily irradiated in early childhood and indicate that this is the first case in the literature The patient was a thirteen year old girl who entered the hospital with the complaint of curvature of the spine which had developed gradually and painlessly over a period of years At the age of six months she had been admitted to the hospital for excision of multiple benign prepuberal melanomata of the back Her entire back was discolored by diffuse nevi within which were numerous discrete coal black melanotic tumors As these tumors are apt to undergo malignant degeneration after puberty they were excised during infancy At the age of nineteen months a large mass was palpable in the left side of the abdomen The mass extended from the costal margin to the level of the iliac crest and beyond the umbilicus medially At that time the spine was straight and the iliac bones were symmetrical A diagnosis of Wilms tumor or embryonal adenomyosarcoma was made and the patient was treated with preoperative irradiation nephrectomy and postoperative

radiation The treatments were given during a period of approximately four weeks through 3 portals: left anterior abdomen, left posterior kidney and left lateral kidney, each field receiving 1100 r in air. The size of the fields varied from 13 × 10.3 to 15.6 × 7.8 cm. One month after treatment, the tumor was excised. No radiation changes were demonstrable in the specimen. About three months later the patient was given postoperative irradiation and this was continued for a period of about three months. The total dose to each field was 3200 r in air. At the age of nine years it was noted that the patient presented a scoliosis in the dorsolumbar region. The right lower extremity was longer than the left, the length of the right femur being 16 inches while the left was 15½ inches. The right tibia measured 13½ inches and the left only 12¾ inches. At the age of thirteen, the scoliosis of the lumbar spine to the right was marked. In addition, there was shortening of the left lower extremity, clubbed foot on the left, and multiple nevi of the skin of the back. Roentgen studies of the lumbar spine showed scoliosis with the concavity toward the left. All of the lumbar vertebrae were uniformly wedged, the left sides of the bodies being diminished in height. The left ilium was smaller than the right. The left twelfth rib was definitely smaller than its fellow on the right. The scoliosis conformed to the expected deformity had the growth of the epiphyses on the left side of the lumbar bodies been retarded. From the history it is obvious that the radiation which the patient received at the age of nineteen months was directed to the left side of the affected vertebrae. The scoliosis did not have the characteristic features expected in idiopathic scoliosis. The inhibition of growth of the left twelfth rib, the left lumbar transverse processes and the left ilium indicate that the effect was due to the x-ray. In the usual types of idiopathic scoliosis, the wedging is maximal at the apex and diminishes gradually towards the ends of the curves, while in the radiation induced curvature the wedging is uniform throughout the affected portion of the lumbar spine.

Interference with the capacity for growth is one of the most important effects of irradiation. Since irradiation in children may produce serious arrest of growth, radiation should not be directed to the epiphyses except for imperative clinical indications. Irradiation of the pelvis and femurs in infants may produce shortening and distortion of the entire extremity and pelvis on the irradiated side and may cause shortening, osteoporosis and necrosis of the femur. Treatment of a malignant tumor of the mouth, neck or tongue may arrest the growth of the mandible and destroy the teeth. While serious but unavoidable effects must be accepted as a necessary consequence, radiation should not be utilized over growing bones in benign conditions.

RADIUM POISONING

Radium is an agent of extremely high toxicity and very small amounts taken into the body produce very profound effects. Waters containing radium are frequently ingested. The toxic agent may gain access to the bones by direct penetration of the mucous membranes of the mouth. Injection of radium solutions may serve as a source of entry. All forms of radium are toxic. The radium is stored in the bones. After implantation in the bones there is a constant irradiation of the marrow, particularly by the alpha rays, with resultant anemia and leucopenia of severe degree. Areas of necrosis appear in the bones

The changes are widespread throughout the skeleton Sequestra form and there is irregular periosteal reaction with proliferation of new bone The skull may be extensively involved, particularly in the regions adjacent to the sutures There is a very high incidence of sarcoma of the osteogenic type in patients suffering from radium poisoning the incidence being so high that a direct relationship doubtless exists The changes in radium intoxication are progressive and permanent The latent period varies widely, one case terminating fatally six years after the initial exposure while others have lived as long as thirty four years after beginning dial painting During the painting of luminous dials the workers formerly pointed the brushes by wetting with the lips The radium salt enters the mouth is swallowed absorbed, and transported to the bones by the blood It is deposited as radium phosphate In adults the radium accumulates in the compacta in amounts greatly in excess of those in the spongiosa which contains red marrow and bone sarcoma results Leucemias have not occurred in these cases Ingestion or injection of as little as 0 01 mgm of radium may cause serious effects in the bones Necrosis of the mandible is not infrequent

ADDITIONAL READING

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Murphy W T and Berrens D L Late Sequela Following Cancericidal Irradiation in Children Radiol *58* 35 1952

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BONE CHANGES IN ELECTRIC CURRENT INJURIES

The widespread generalized use of electricity in industry and the household has resulted in an increase in the number of electric current injuries and burns *In 1924 Palugyay called attention to the bone* lesions which occur after electric burns and described the roentgen manifestations in two cases The first bone change subsequent to electric burns or injuries comprises the development of delicate zig zag lines in the bones and osteoporosis of the affected extremity Subsequently there ensue sclerosis hypertrophy of the periosteum and alterations in the contour of the bone In some instances there may be a clear cut line of demarcation between the lesion and the unaffected portions of the bone Sequestrations may form Osteoporosis usually affects all the bones of the injured extremity and marked atrophy may be demonstrable roentgenographically within a few days after the trauma In association with the atrophy sclerosis may ensue in the bones not immediatly adjacent to the soft tissues affected by the electric current At first, the sclerosis conforms to the shaft of the bone With the passage of time the sclerotic area becomes separated by a narrow band of increased radiance indicating that the periosteum is separated and elevated Extreme atrophy of the bone in the injured extremity is the striking roentgen finding and in many instances comprises the sole manifestation There may be softening of the osseous structures leading to contractural deformities of the extremity Pathologic fractures may occur The porosis of the affected portion of the skeleton is similar to that in Sudeck s atrophy Palugyay was not certain whether the periosteal sclerosis repre

sented an inflammatory reaction, resulted from a direct injury to the cells of the periosteum with subsequent ossification, or was due to subperiosteal hemorrhage. Jaffe is of the opinion that the extensive bone changes are due not only to the initial electric shock and burn but also to lesions which develop in the blood vessels and the nerves.

ADDITIONAL READING

Jaffe, R. H.: Electropathology: Review of Pathologic Changes Produced by Electric Currents. Arch. Path., 5: 837, 1928.

L[illegible], J.: Knochenveränderungen Bei Verletzung Durch Elektrischen Strom. Fort. a. d. Geb. d. Rönt., 32: 568, 1924.

Chapter

7

Diseases of the Joints and Periarticular Tissues

THE rheumatic diseases have been divided into various groups by the American Rheumatism Association. The classification suggested by the Association will, with slight modification, be used in the following discussion.

ARTHRITIS DUE TO INFECTION. INFECTIOUS ARTHRITIS

In the arthritides due to infection are included the diseases of the joints that result from infection with the staphylococcus, gonococcus, streptococcus, brucella, meningococcus, treponema pallidum, pneumococcus, tubercle bacillus, and other less common microbial agents.

Acute Pyogenic Arthritis

Arthritides due to the gonococcus, meningococcus, pneumococcus, staphylococcus and streptococcus, the pyogenic cocci, are similar in their manifestations. The staphylococcus and hemolytic streptococcus are the etiologic agents in the majority of cases. The arthritic process varies with the severity and duration of the infection. The synovial tissue becomes hyperemic, swollen, thickened and infiltrated with cells that are predominantly polymorphonuclear leukocytes. The articular cartilage may be irreparably destroyed with resultant ankylosis. The causative microorganism may be demonstrated by blood cultures, or bacteriological examination of aspirated synovial fluid establishes the diagnosis with definiteness.

The articular complications of gonorrhea usually appear within ten to thirty days after the initial infection. There are no characteristic roentgen changes which distinguish it from other types of suppurative joint disease. Damage to the articular cartilage and the adjacent bone is evidenced by osteoporosis, periostitis, osteomyelitis or epiphysitis. Fluid in the joint is manifested by widening of the joint space. The periarticular tissues are increased in width and density and the outlines of the muscle bundles are blurred. Soft tissue x-ray studies are invaluable in the establishment of the diagnosis. A characteristic manifestation is destruction of the articular cartilage with narrowing or obliteration of the joint space. The destruction involves the entire cartilage. The changes develop rapidly in the case of a virulent process and may become very extensive within a few weeks. The alterations are most marked and develop first at the points of weight bearing. The articular surfaces become destroyed and there is roughening and irregularity of outline of the adjacent bony surfaces. Pathologic dislocation may ensue. Advanced osteoporosis develops in the bones about the affected articulation.

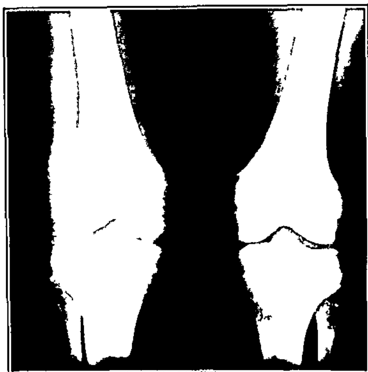


Fig 227 Arthritis of the Knee Due to Gonococcic Infection The right knee joint space is slightly widened and the bones show a mild degree of osteoporosis The soft tissues about the right knee are thickened and swollen The changes are those of early infectious arthritis



Fig 228 Arthritis of the Knee Due to Tuberculosis Early Stages *A* Anteroposterior view There is marked soft tissue swelling about the knee and lower femur on the right The knee joint space is narrowed and irregular and there is a marked degree of osteoporosis *B* Lateral projection The soft tissue swelling osteoporosis and joint changes are well illustrated

Tuberculous Arthritis

Tuberculosis of bones and joints occurs in approximately 4 per cent of patients with tuberculosis. In pulmonary tuberculosis a primary tuberculous lesion develops in the lung or pleura as a result of infection through inhalation. The bacilli spread to the bronchopulmonary lymph nodes and hematogenous dissemination results, the organisms metastasizing to bone or synovial tissue. Bacilli may travel through lymph channels from the pleura to the vertebral column. Arthritis may also develop by penetration into the joint from adjacent diseased bone. The urogenital tract, the liver and the spleen also serve as primary sources, caseous foci in these regions frequently causing hematogenous dissemination. The disease is most common in children although it may occur at any age. Negroes are especially susceptible.

The joints of the lower limbs are involved much more frequently than those of the upper. The spine is affected oftener than any peripheral joint, the bodies of the lower thoracic and upper lumbar vertebrae being the commonest site of infection. As caseation ensues, the bone loses strength and collapses and spread occurs from the area. In the case of the spine, the disease extends along the anterior or posterior longitudinal ligaments and may involve several contiguous vertebrae both proximally and distally. The infection may spread from one vertebra to the next through the intervertebral disc. The disease is characterized by caseation and abscess formation. The abscess usually dissects between fascial planes or muscle bundles and may produce a discharging sinus at a distant point. The infection may remain quiescent and become arrested with resolution and healing by fibrosis or bony fusion. Granulation tissue forms in the joint and undermines and separates the articular cartilage. Periarticular swelling develops with relatively little effusion. The articular cortex and neighboring bones are often destroyed.

Roentgen Manifestations. The early signs consist of osteoporosis, periarticular swelling, synovial effusion and thickening. Decalcification of bone increases and marginal erosion with thinning and interruption of the articular cortex follow. The joint space is preserved until late, but in the advanced stages sequestration, abscess and sinus formation may become evidenced as bone destruction progresses. Calcium deposition and new bone formation are absent. In tuberculosis of the knee, shoulder, and similar joints of the extremities, the articular cartilage is protected at the points of contact and pressure of opposing articular surfaces and the destruction occurs peripherally along the free aspects of the bones in regions in which the tuberculous granulations are able to grow. The proteolytic ferments which are present in infectious arthritis are absent in tuberculous arthritis and the masses of dead cartilage may persist with relatively little progression of the destruction. Disappearance of the bony cortical shadow is a manifestation of bone destruction and usually begins peripherally at the margins of the weight bearing portions of the joint surfaces in the region in which there has been absorption by granulation tissue. There may be preservation of the joint space for periods of months or years. In pyogenic arthritis the articular cartilage undergoes destruction at the points of contact and pressure of opposing articular surface. Acute suppurative arthritis is characterized by severe osteoporosis in the region of the joint, while advanced osteoporosis is usually lacking in tuberculosis. Acute pyogenic joints manifest a distinct tendency to repair and ankylosis whereas these features are customarily absent in tuberculosis.



Fig 229 Tuberculosis of the Knee Tuberculosis of the knee characteristically produces a rounded sharply defined area of bone destruction in the condyle of the tibia adjacent to the articular surface The patient a man of thirty years had been under treatment for pulmonary tuberculosis during the past four years



Fig 230 Arthritis of the Shoulder Due to Tuberculosis There is an extensive area of rarefaction and irregularity of outline involving the head tuberosity and upper shaft of the humerus The shoulder joint space is narrowed There is marked osteoporosis The soft tissues of the left shoulder are decreased in size and density The patient had advanced bilateral pulmonary tuberculosis

There are atypical cases termed caries sicca. This type of tuberculosis is characterized by the formation of multiple rounded, punched out, bony erosions and excavations in the region of the joint. The cartilage may be preserved. There is no evidence of repair and little or no soft tissue reaction.

Tuberculosis of the Hip

In tuberculosis of the hip the earliest manifestations are atrophy of the muscular tissues and the bones about the joint. This is manifested by a decrease in the size of the soft tissues and increased radiance of the bones. The joint space is narrowed or, in the presence of fluid, may be slightly increased in width. The contours of the bone become hazy and indistinct. There is deformity of the articular ends of the affected bones and scattered areas of rarefaction of varying size appear in the bone. The capsule is distended and thickened. An early roentgen sign of tuberculosis of the hip in young individuals is slight lateral displacement of the head of the femur with widening of the joint space. Beaking of the upper edge of the acetabular rim occurs in cases in which the infection originates in the acetabulum. The beak in some instances becomes elongated and resembles a bone graft extending from the neck of the femur to the innominate bone. Sequestrum formation is rare in young patients and usually is a manifestation of secondary infection. In adults it is relatively common and is indicative of a poor prognosis. Early diagnosis is of the utmost importance as prompt institution of therapy may prevent destruction of the joint, ankylosis, interference with growth and other sequelæ.

Tuberculosis of the Spine. Pott's Disease

The classical roentgen appearance of Pott's disease comprises narrowing of the intervertebral discs, extensive destruction of the vertebral bodies and in many instances the shadow of a cold abscess. The changes are specific and as a rule permit of a definite diagnosis. There are many variations in the manifestations and the characteristic changes are frequently not present. The disease may occur at any age from early infancy to the seventh and eighth decades of life. The most common period is in the second and third decades. In many instances the disease is relatively benign and undergoes healing. Those that do not heal may progress slowly and persist for many years. It is unusual for the disease to be limited to one vertebra. When multiple vertebræ are affected, the process may involve contiguous vertebræ or appear simultaneously in different regions. The lower dorsal and lumbar vertebræ are the most frequently involved. The disease originates in the vertebral body and the intervertebral disc is involved by direct extension. The reason for this probably lies in the anatomical character of the intervertebral joint spaces, as these structures have no capsule or synovial lining and are relatively avascular. The lesion usually is in the anterior aspect of the vertebral body, being less common in the central and posterior portions. The process may occur in two forms, a productive or sclerotic type and an exudative caseous or destructive variety. In some instances both are present simultaneously. Usually the exudative type is associated with productive changes at the periphery of the caseous area. This variety is more common and comprises the classical picture of the disease. Areas



Fig 231   Fig 232

Fig 231 Tuberculosis of the Cervical Spine There is destruction of the anterosuperior portion of the fifth cervical vertebra The intervertebral disc between the fourth and fifth cervical vertebrae is narrowed and there is kyphosis at this level The soft tissues of the prevertebral region are increased in density and thickness

Fig 232 Tuberculosis of the Spine There are advanced destructive changes involving the bodies of the twelfth dorsal and first lumbar vertebræ The intervertebral space between these vertebræ is obliterated There is kyphosis at the level of the lesion



Fig 233   Fig 234

Fig 233 Tuberculosis of the Spine Calcified Abscess There is tuberculosis of the dorsolumbar spine with extensive destruction of the bodies of the twelfth dorsal and first lumbar vertebræ The intervertebral spaces between the eleventh and twelfth dorsal vertebræ and the twelfth dorsal and the first lumbar vertebræ are obliterated There is bilateral calcification in the paraspinal region The calcific deposits extend downward and laterally along the margins of the first and second lumbar vertebræ (arrow)

Fig 234 Tuberculosis of the Spine with Severe Kyphosis

of rarefaction of varying size develop within the affected zone These areas are composed of gelatinous material and frequently contain bony fragments Progression of the process results in extension to the intervertebral disc Compression of the vertebra ensues with the formation of a gibbus Sequestrum may be produced as a caseous process encircles normal bone or bone which has undergone productive changes The productive form presents tuberculous granulation tissue which is rich in capillaries and fills the marrow space The granulation tissue erodes the bony trabeculæ



FIG 235 Tuberculosis of the Spine There is irregularity of outline and eburnation involving the adjacent portions of the bodies of the first and second lumbar vertebræ There is bony bridging between these vertebræ and the intervertebral disc is narrowed

A process limited to the central portion of the vertebra is difficult to diagnose as pyogenic or fungus infection may produce a closely similar picture In the productive form of the disease the affected vertebræ become densely sclerotic

In many instances the lesion is associated with a tuberculous abscess of the spine The abscess forms a soft tissue mass which is spindle or fusiform in shape It is usually symmetrical and may present slight irregularity of its outer margins The presence of an abscess may be the first roentgen sign of tuberculosis and in some instances precedes visible changes in the vertebral bodies Bilateral paravertebral abscess is the most characteristic feature of tuberculosis of the spine In rare cases the abscess may be unilateral The abscess may become calcinated The presence of areas of mottled calcification in the paravertebral region in association with changes in the vertebræ and the intervertebral discs is

characteristic of tuberculosis of the spine and establishes the diagnosis with a high degree of certainty in most instances. A significant complication is tuberculosis of the adjacent ribs. This may furnish a clue to the presence of vertebral disease. One or more ribs may be involved. In the dorsal region a valuable thoracic paravertebral landmark is formed by the postero-mesial pleural line. This linear shadow lies to the left of the vertebral column and represents the postero-mesial boundary of the left lung. It is only rarely seen on the right. Displacement of the linear shadow by a left-sided unilateral tuberculous abscess constitutes an important diagnostic finding.

Confusion of a tuberculous abscess with a spinal neurofibroma is unlikely. Most neurofibromas are dumbbell shaped. The tumor arises from the posterior nerve roots and is composed of a spinal portion within the vertebral canal and a larger segment lying extrapleurally and paravertebrally. The two portions of the lesion are joined by a narrow neck which passes through the intervertebral foramen. The tumors are usually located in the upper and posterior mediastinum. The roentgen manifestations are characteristic, there being a rounded, sharply defined shadow of soft tissue density projecting to one side of the spine. There may be erosion of the adjacent rib margins. Enlargement of the intervertebral foramen is an important sign. There may be increase in the width of the intercostal space at the level of the lesion. This is thought to be due to pressure exerted by the expanding tumor. In the presence of a unilateral paravertebral abscess and adjacent bone destruction, the diagnosis of tuberculosis can be established with ease. However, in a unilateral tuberculous abscess without characteristic changes in the adjacent bone, an erroneous diagnosis of neurofibroma is often made.

Tuberculosis of the Sacro-iliac Joints

Tuberculosis of the sacro-iliac joints is not rare. The roentgen manifestations are characteristic and in many instances permit of a definite diagnosis. In the early stages there is haziness or loss of definition of the joint outlines. Later there develop irregularity of the articular surfaces and areas of erosion. If the disease subsides at this stage the joint space becomes narrowed and the adjacent bone shows increased density. With progression of the infection the bone destruction becomes more marked. Bony ankylosis develops in the late stages. In some instances there is increased density about the margins of the joint without bony fusion. Draining sinuses frequently form. The disease is usually associated with tuberculosis of the hip and other parts of the body.

Syphilitic Arthritis

The incidence of syphilitic joint disease has been declining steadily because of earlier more effective antisyphilitic therapy. Arthritis may occur at any age as a complication of either the congenital or acquired disease and may result from direct spirochetal infection of the joints or from neurogenic changes (Charcot's joints) secondary to tabes. The latter constitute about half of the patients in the acquired group. The joint complications of syphilis may manifest themselves as arthralgia (early acute syphilis), osteochondritis, epiphysitis, periostitis, painless,

of rarefaction of varying size develop within the affected zone These areas are composed of gelatinous material and frequently contain bony fragments Progression of the process results in extension to the inter vertebral disc Compression of the vertebra ensues with the formation of a gibbus Sequestrum may be produced as a caseous process encircles normal bone or bone which has undergone productive changes The productive form presents tuberculous granulation tissue which is rich in capillaries and fills the marrow space The granulation tissue erodes the bony trabeculae



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RHEUMATIC FEVER

Rheumatic fever is an acute or chronic inflammatory process initiated by a preceding group A hemolytic streptococcic infection. The inflammation is disseminated in the connective tissues of many organs. It occurs at all ages, with the peak incidence between six and nine years of age, but there is an appreciable incidence in adult life. The disease is characterized by polyarticular migratory joint involvement. Roentgen examination reveals osteoporosis about the involved joints in many instances. Not infrequently, there are no roentgen manifestations, the joints presenting an entirely normal appearance.



FIG 238



FIG 239

FIG 238 Rheumatoid Arthritis of the Shoulder. The articular surface of the humerus and the glenoid are markedly irregular in outline. The shoulder joint space is narrowed. There is advanced osteoporosis.

FIG 239 Rheumatoid Arthritis Involving the Sacro iliac Joints. There is narrowing, haziness and irregularity of outline of the sacro iliac joints with marked eburnation about the margins of the joints.

RHEUMATOID ARTHRITIS, ATROPHIC ARTHRITIS, PROLIFERATIVE ARTHRITIS, POLYARTHRITIS, CHRONIC INFECTIOUS ARTHRITIS

Rheumatoid arthritis is a disease of unknown cause. Eighty per cent of the cases occur between the ages of twenty five to fifty, the highest incidence being in the third decade. The disease affects primarily the interfibrillar substance of the connective tissue. In the early stages there is thickening of the synovial lining. As the disease progresses, the articular cartilage may ulcerate and be destroyed. Fibrous or bony ankylosis ensues. The disease usually begins insidiously. One or more joints gradually become swollen and painful and others soon are affected. The joints of the feet may be the first involved, the characteristic changes

symmetrical synovitis (Clutton's joints in eight to sixteen year old children, frequently with interstitial keratitis), gummatous lesions, or Charcot's joints A positive serologic test for syphilis with synovial fluid and/or blood is not conclusive evidence of syphilitic arthritis The diagnosis is presumptive in the presence of (1) positive serologic test for syphilis, (2) other stigmas of syphilis, and (3) characteristic changes in the bones as seen in roentgenograms Charcot s disease is discussed in the section entitled neurogenic arthropathy (see p 496)

Fig 236



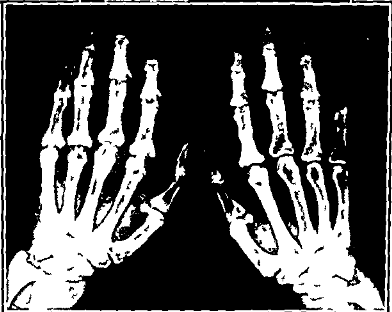


Fig 237

Fig 236 Rheumatoid Arthritis There are extensive destructive changes involving the joints of the wrist and hand There is osteoporosis of the hand and the wrist The metacarpophalangeal joints of the index and middle fingers are partially dislocated

Fig 237 Rheumatoid Arthritis There is extensive destruction involving the joints of the fingers and hands The process is more advanced on the right There is marked osteoporosis

and atrophy and cutaneous changes are usually absent the erythrocyte sedimentation rate is not rapid, and osteoporosis and ankylosis do not ensue Differential diagnosis between adult rheumatic fever and rheumatoid arthritis is difficult The arthritis of atypical rheumatoid arthritis is apt to be progressive rather than migratory, while in adult rheumatic fever it is either progressive or migratory The progression of the arthritis may be slower in rheumatoid disease than in rheumatic fever The therapeutic ineffectiveness of salicylates with failure to halt progression characterizes their action in rheumatoid arthritis In rheumatic fever the objective improvement is dramatic after the proper administration of the salicylates and the medication exerts an equally effective prophylaxis against recrudescences of frank arthritis Roentgen studies in rheumatic fever except for transient osteoporosis are essentially negative

Juvenile Rheumatoid Arthritis Still's Disease

Still's disease is a systemic condition The arthritis in adults is the most obvious and important manifestation while the bone and joint disturbances in children are less striking and may appear only later in the course of the disease The juvenile form of the disease first described by Still in 1897, is a special clinical form of rheumatoid arthritis The onset may be sudden with joint pains, fever, rash leukocytosis anemia and possibly pericarditis Later there is adenitis and splenomegaly In some instances there is an insidious onset with gradually increasing joint pains and swellings as in the adult forms of the disease Psoriasis may occur as an associated lesion The condition usually begins between the ages of two to five years The sex distribution is equal In children affected before the age of six years, the mortality may be as high as 30 per cent On histopathologic examination the muscles show atrophy with small focal perivascular accumulations of lymphocytes and plasma cells There are often granulomatous subcutaneous nodules over the ulna and sacrum Adhesive pericarditis is frequently present In the joints, the changes comprise marked proliferation of the synovial membrane, hypertrophy of the connective tissue in the marrow spaces of the subchondral bone and vascularization and fibrosis of the joint capsule The synovial tissue undergoes proliferation grows over the articular cartilage and invades and becomes adherent to the cartilage Extensive destruction of cartilage ensues and the fibrous adhesions ossify with resultant bony ankylosis

Roentgen Manifestations The roentgen changes are divisible into four principal groups (1) There is a periosteal change which commonly affects the entire length of a metacarpal or metatarsal and involves the medial or lateral aspect of the bone but not both surfaces The periosteal reaction may resolve completely leaving no trace or may persist as cortical thickening or a periosteal tab of bone (2) The articular cartilage may be destroyed partially or completely (3) The effect on bone usually comprises porosis The porosis may be slight, moderate or marked and often is associated with bone destruction due to invasion of the granulations from the synovia The destruction may undergo arrest at any stage with return to practically normal bony density However, there may be no formation of new bone trabeculae Progressive and severe destruction may result in subluxation of the joint or bony ankylosis (4) There may

in the hands appearing later In the typical case the articular involvement is symmetrical The smaller joints of the hands are prone to be affected, especially the proximal interphalangeal joints of the fingers The terminal interphalangeal joints usually escape, an important aid in the differentiation from degenerative joint diseases In severe cases almost every joint in the body may become involved There is periarticular swelling and pain with limitation of motion The joint presents a spindle shaped appearance The bursas and tendon sheaths are frequently involved much of the swelling being due to synovial proliferation and edema Involuntary muscle splinting occurs with a resultant contraction usually in flexion Fibrous and bony ankylosis accompanied by subluxation or dislocation develop in the affected joints Atrophy is particularly marked in the muscles of the hands and contributes to the fusiform appearance of the joints The muscle atrophy is an integral part of the disease and is not attributable to disuse and muscular malnutrition associated with persistent contraction Joint changes indistinguishable from rheumatoid arthritis are present in some patients with ulcerative colitis Splenic enlargement occurs in about 10 per cent of patients with the disease Generalized lymphadenopathy of moderate degree is commonly observed The presence of previous or concurrent rheumatic fever must be considered Iritis and uveitis, often of an intractable nature may accompany or precede rheumatoid arthritis and may lead to blindness There is usually a normocytic hypochromic anemia of the iron deficiency type The white blood cell count the differential count and the platelets are within normal limits in most patients The erythrocyte sedimentation rate is usually rapid Group A hemolytic streptococci are agglutinated by the serums of a considerable number of patients with peripheral rheumatoid arthritis

Roentgen Manifestations In the incipient stages there may be no demonstrable abnormalities Intracapsular effusion, periarticular soft tissue swelling and generalized demineralization occur in the region of the affected articulations There are spindle shaped swellings of the soft tissues about the affected joints and slight increase in the width of the joint space There may be narrowing of joint spaces and marginal resorption with mottling of bone Cortical destruction of the metatarsal heads may appear early As the disease progresses there develops narrowing of the joint space due to destruction of cartilage Small areas of cortical erosion appear as 'punched out' areas and are a prominent feature of rheumatoid arthritis and gout In advanced cases the articular surfaces become fused There may be dislocation with destruction of the ends of the bones especially in the fingers Varying degrees of bone production lipping and osteophyte formation may occur and are evidences of degenerative joint disease secondary to the rheumatoid arthritis Marked osteoporosis ensues In the late stages of the quiescent form of the disease there is re establishment of practically normal contrast between the spongiosa and compact bone or there may be reconstructive changes with cystic enclosures and increased density of the bones in the periarticular regions and the shafts

Differential Diagnosis The differentiation between rheumatoid arthritis and degenerative joint disease is important as there is a great difference *in prognosis and treatment Rheumatoid arthritis is a systemic* disease and the patients are ill In degenerative joint disease the patients are usually well save for their joint complaints muscular weakness

occur simultaneously with eburnation in different parts of the same joint indicating that ankylosis and eburnation represent different phases of the same pathological conditions Marginal osteophytes may occur in quiescent or arrested stages of rheumatoid arthritis of the peripheral joints and the adjacent bones usually become rarefied, the degree of rarefaction being proportional to the activity and duration of the lesion There is rarefaction of the vertebral bodies in the advanced stages the change corresponding to the para articular rarefaction in other joints Pain and limitation of motion are usually present when the apophyseal joints are involved Swelling cannot be recognized in the case of the spine because of the inaccessible location of the joints but can be inferred from the associated radicular neuritis caused by compression of the nerve roots



Fig 240

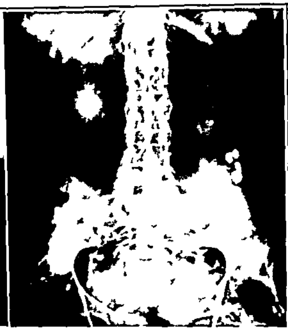


Fig 241

Fig 240 Rheumatoid Arthritis Marie Strumpell Type There is complete bony bridging along the margins of the bodies of the vertebræ The spine shows advanced osteoporosis

Fig 241 Marie Strumpell Arthritis There is bony bridging along the lateral margins of the dorsal and lumbar vertebræ The sacro iliac and hip joints are completely obliterated There is generalized porosis of the bones of the spine pelvis and hips

Spurs of the Calcaneus in Marie Strumpell Arthritis Spurs on the plantar surface of the calcaneus in young adults may occur in association with various toxemias In men between the ages of eighteen and thirty with manifestations of periostitis or spurs of the calcaneus, there are frequently associated changes in the lumbosacral spine and the sacro iliac joints characteristic of Marie Strumpell disease The pathogenesis is not clear It is believed that in the acute phase of Marie Strumpell disease there develops a periostitis of the calcaneus which progresses to form a spur the spur formation at the attachment of the plantar fascia to the anterior aspect of the tubercle of the calcaneus being similar to the bony prolifera tion which occurs in other locations in this disease Roentgen therapy relieves the pain associated with spurs of the calcaneus

be acceleration of growth of the epiphysis or the ossific centers such as the carpal bones. Premature fusion of the epiphyses may occur with gross deformities as the result of fusion of one epiphysis while the growth of an adjacent epiphysis continues. The juxta epiphyseal parts of the long bones may taper, the effect being similar to the destruction of the ends of the long bones by synovial proliferation in adults. The joints most frequently affected are the wrists, feet, ankles, knees and cervical spine.

Felty's Syndrome

The term Felty's syndrome has been applied to patients with rheumatoid arthritis and hepatomegaly, splenomegaly, and leukopenia. These patients usually have severe disease. This syndrome has been called rheumatoid arthritis with hypersplenism.

Rheumatoid Arthritis of the Spine. Marie-Strumpell Arthritis. Von Bechterew Spondylitis

The syndrome known as ankylosing arthritis, Marie Strumpell arthritis, spondylitis ankylopoietica, and spondylitis rhizomelique comprises rheumatoid arthritis of the spine. About 20 per cent of the patients have changes in the peripheral joints indistinguishable from those in classical rheumatoid arthritis and this syndrome may merely be a special form of the latter disease. There are many dissimilar features. The sex ratio is nine or more males to one female, in sharp contrast to typical rheumatoid arthritis. The results of the hemolytic streptococcus agglutination and sensitized sheep cell tests are almost always negative in patients with spondylitis. Roentgen therapy causes a symptomatic remission in over 80 per cent of patients with rheumatoid spondylitis. There is decrease in pain and stiffness and increase in mobility of the spine. Although the patients have relatively few subjective symptoms of pain and stiffness, the roentgen changes and increasing rigidity of the spine continue to progress. Roentgen therapy has proven effective in all stages of the disease and in some instances produces regression of the changes.

Roentgen Manifestations. There is a marked degree of osteoporosis with no collapse or wedging of the vertebrae. The intervertebral spaces are normal in width or slightly narrowed. There is marked calcification of the ligaments along the margins of the spine and in the spaces between the bodies of the vertebrae. The sacro iliac joints are markedly narrowed, irregular in outline, hazy and may be ankylosed. The changes often begin in the lower spine, are rapidly progressive and the entire spine is usually involved. The costovertebral joints are narrowed or obliterated. The manifestations are localized in the apophyseal joints which connect the contiguous articular processes. These joints are a synovial or diarthrodial articulation similar to the joints of the limbs and must be differentiated from the vertebral symphysis which is the articulation between two vertebral bodies and the intervening vertebral disc and comprises a synchondrosis or symphysis similar to the symphysis pubis. *Rarefaction of the articular* processes with narrowing of the intervening intervertebral space is present in the incipient and early stages. The disease may progress rapidly and continuously to complete bony ankylosis or may advance slowly. There is roughening and eburnation of the articular surfaces. Anklyosis may

occur simultaneously with eburnation in different parts of the same joint indicating that ankylosis and eburnation represent different phases of the same pathological conditions Marginal osteophytes may occur in quiescent or arrested stages of rheumatoid arthritis of the peripheral joints and the adjacent bones usually become rarefied the degree of rarefaction being proportional to the activity and duration of the lesion There is rarefaction of the vertebral bodies in the advanced stages the change corresponding to the para articular rarefaction in other joints Pain and limitation of motion are usually present when the apophyseal joints are involved Swelling cannot be recognized in the case of the spine because of the inaccessible location of the joints but can be inferred from the associated radicular neuritis caused by compression of the nerve roots



Fig 240 Fig 241

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TRAUMATIC ARTHRITIS

Injury to a joint may result in hemorrhage or the formation of an exudate with distension of the joint The joints most commonly affected are the knee, ankle, elbow, shoulder, and hip, although any articulation of the body may be affected Injury to the cartilage and synovia may be followed by the formation of a scar with resultant interference with function and permanent alterations in the joint The causative factor may be a single severe blow, repeated slight traumas or gunshot wounds needles, missiles projectiles, and other foreign bodies A fracture involving the joint, an epiphyseal separation, or a dislocation may cause traumatic arthritis Dislocation of the semilunar cartilage is frequently followed by chronic arthritic changes Fractures involving a joint often result in the formation of marginal osteophytes In simple fractures, the callus is usually endosteal in character, although periosteal callus may form along the margins of the bones The amounts of callus are greatly increased in young children With separation of the epiphysis, prompt reduction is essential in order to prevent subsequent deformity and interference with growth It must be stressed that foreign bodies particularly those of the nonopaque variety, may remain in the joints for long periods of time The foreign bodies result in degeneration of the articular cartilage hypertrophy of the synovial membrane, and fibrosis of the periarticular tissue These changes may be followed by the characteristic manifestations of degenerative arthritis Soft tissue structures in and about the joints such as the capsule and synovia may become separated and after the passage of months or years, undergo calcification with the formation of joint mice Trauma may precipitate other forms of arthritis, particularly the tuberculosis, pyogenic, and gouty forms of the disease Any form of arthritis, especially degenerative joint disease, may be aggravated following trauma The changes in traumatic arthritis comprise a synovitis with the formation of an effusion The joint space becomes widened and the periarticular soft tissues are widened and increased in density The presence of a fracture or foreign body should be carefully sought for in every instance In the chronic stages roentgen manifestations may be absent or comprise osteoporosis osteophyte formations or other changes associated with the various forms of arthritis

NEUROGENIC ARTHROPATHY CHARCOT'S DISEASE

Any disease impairing sensation particularly proprioceptive sensation in joint structures predisposes the joint to traumatic damage and may result in severe deformity of the joint The Charcot joint of tabes dorsalis is the typical example of this Other disease processes such as syringomyelia, diabetes mellitus leprosy transection of the spinal cord diabetic neuropathy and peripheral nerve injuries may lead to similar arthropathies In tabes dorsalis there occur very marked changes with painless chronic swelling involving one or several joints The spine is particularly apt to be affected In lues the condition is a manifestation of tertiary syphilis and is due to disturbance of the trophic nerves with loss of the deep pain sense The motor nerves are unaffected Ataxia is present The involved joints are markedly swollen relaxed and show abnormal mobility There is no pain on motion or weight bearing Crepitation

may be present. The disease is chronic and progressive. Practically any joint may be involved. Due to loss of the pain sense and the development of ataxia the joints undergo increased wear and tear and the ligaments and capsule become weakened, stretched and torn.

On roentgen examination there is extensive destruction of the joint, in some cases it being impossible to identify the component parts of the articulation. The periarticular soft tissues are swollen and dense. Large masses of calcification are present in the soft tissues about the joint. The bony masses vary widely in size and density and may be smooth or irregular in outlines. There is marked sclerosis of the ends of the bone with absence of osteoporosis or muscle atrophy despite the presence of extensive destructive changes. Subperiosteal proliferation takes place in regions in which the periosteum is elevated adjacent to the areas of destruction. Osteoporosis may ensue late in the disease after the paralytic

Fig. 242

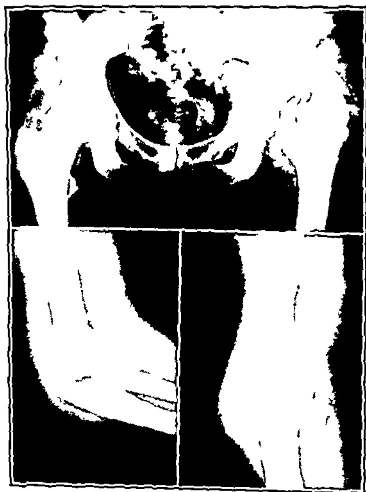


Fig. 243

Fig. 242. Charcot's Disease. There is destruction of the head and neck of the left femur with upward and inward displacement of the shaft. There are multiple fragments of bone of varying size and shape in the soft tissues about the hip. There is no evidence of atrophy of the bones of the left side of the pelvis or the femur.

Fig. 243. Charcot's Disease of the Elbow. There is complete destruction of the elbow joint. There are multiple large irregular fragments of bone scattered in the soft tissues. There is sclerosis of the bones of the elbow.

stage has developed The articular cartilage is eroded the adjacent cortex is destroyed, and disintegration of the joint results Spurs develop at the ends of the bones and may remain attached to the bone or separate to be free in the soft tissues Fractures may occur and frequently fail to unite Secondary infection may cause osteomyelitis or arthritic changes



Fig 244 Neurogenic Arthropathy Syringomyelia of the Shoulder There is absorption and fragmentation of the upper end of the humerus The shoulder joint is destroyed

VERTEBRAL NEUROGENIC ARTHROPATHY CHARCOT'S DISEASE OF THE SPINE

Charcot s disease of the spine occurs in about 3 to 10 per cent of patients with tabes, being more common than is usually believed The condition occurs most commonly in the fifth and sixth decades of life and may develop fairly rapidly It is more frequent in men than in women, probably due to the greater incidence of trauma in the male Clinical manifestations of tabes dorsalis may not be present and the serological tests are not positive in every instance The common site is the lumbar region the second to fifth lumbar vertebræ usually being involved The lower thoracic spine is less frequently affected and the cervical spine is involved but rarely The lesion may be localized to a single vertebra although this is not the rule There is kyphosis or kyphoscoliosis of the lumbar or dorsolumbar spine The alterations caused by the deformities of the vertebral bodies and the new bone formation may be palpable Tenderness and pain are absent The movements of the spine are excessively free because of the hypotonia of tabes despite the fact that the

affected segments are rigid A sharp, short thud on flexion and extension movement of the spine is a characteristic finding Antiluetic therapy is of no value Spinal fusion may be necessary in spinal osteoarthropathy Support by a brace or corset is an important therapeutic measure and tends to limit the deformity

Roentgen Manifestations The roentgen manifestations comprise extensive disorganization and destruction associated with massive new bone



Fig 245 Neurogenic Arthropathy The articular surface of the tibia is destroyed and the knee joint space is obliterated There are multiple large fragments of bone in the region of the tibial condyles The bones are markedly porotic

formation Lateral subluxation of the vertebral body frequently develops The new bone formation occurs as large, beaked osteophytes or as massive paravertebral new bone The formations of new bone may lead to ankylosis The disc spaces are narrowed The transverse processes may be involved in both the destructive and proliferative processes Sclerosis of the bone in the region adjacent to the articular cortex is an important diagnostic sign Osteoarthritic spurs may be present Other joints may be affected simultaneously, the larger joints frequently being involved, particularly the hip and the shoulder

Differential Diagnosis In osteoarthritis, the spur formations are less marked and the progression of the changes is less rapid There is pain and limitation of motion Gross destruction and disorganization are not present Syphilitic infection of the lumbar spine is rare It is manifested as a diffuse osteitis with thickening and hardening of adjacent vertebrae or as gummatous formations with extensive destruction of the vertebrae Tuberculosis does not present new bone formation or sclerosis and is usually associated with pain and limitation of motion In calcification of a paravertebral group of lymph nodes the changes may simulate massive new bone formation, however, the vertebrae are not affected A previous fracture with wedging, the so called Kummell's disease, may cause difficulty in diagnosis There is usually a history of a previous trauma the changes are less marked, and successive roentgen studies at intervals show that the lesion is not rapidly progressive

Joint Changes in Paraplegia Paraplegic Neuroarthropathy

Paraplegic neuroarthropathy is particularly apt to occur in paralysis of the lower half of the body subsequent to disease or injury of the spinal cord The condition is associated with two principal types of changes (1) osteoarthropathy and (2) soft tissue calcification in the regions of the joints The joint changes vary from slight periarticular rarefaction and narrowing of the joint spaces to practically complete destruction of the articulation Subluxations are common There is marked osteo porosis Osteoarthropathy of the Charcot type may occur rarely The spine is usually devoid of abnormalities except those due to the original injury such as fracture of the vertebrae The lesions in the joints may develop within a few months to several years after the injury There is no correlation between the duration of the disability and the extent of the changes In some instances patients with injuries sustained less than a year previously show very extensive lesions while others present no changes twenty years following the injury

Soft Tissue Calcifications in Cord Lesions

The first description of soft tissue calcifications in cord lesions was published in 1919 by Dejerine and her co workers and comprised an extensive study of the clinical anatomical and pathological aspects of soft tissue calcification The calcific depositions develop below the level of the cord lesion and may be single or multiple The most common sites are in the vicinity of the hip joints the medial condyles of the femurs and the knee joints The calcifications usually show rapid progression The processes occur in areas where no previous hemorrhages or infections have taken place In some instances there develops a lamellated formation of bone about the joint The calcific depositions may surround the articular capsule and markedly limit the motion of the adjacent joints The calcification may appear relatively early after the injury, authenticated cases having been described within twenty days after the trauma The patients show a high degree of calculi in the urinary tract and these also may develop very rapidly after the injury The changes may occur simultaneously in more than one area While the calcifications may develop within twenty days after an injury, there are other instances in which they have developed in individuals who have been paralyzed for ten fifteen or twenty years

Patients with soft tissue calcifications may suffer fractures below the site of the calcification. The fractures occur after minimal trauma or without apparent trauma. This is most probably due to osteoporosis; hence the fractures are classified as pathological fractures. The patients do not have any pain below the level of the cord lesion and the swelling or deformity associated with the fracture may be the first manifestation of the break of the bone. In some instances the fracture is discovered as an incidental finding. The fractures show an unusual reaction during healing. There is exuberant callus formation which is greatly excessive in amount. The callus may be so extensive as to simulate a subperiosteal hemorrhage and lead to a mistaken diagnosis of scurvy. The condition



Fig. 246. Joint Changes Associated with Paraplegia. There is extensive destruction of the pubis, ischium, acetabulum and the head and neck of the femur. The femur is displaced upward.

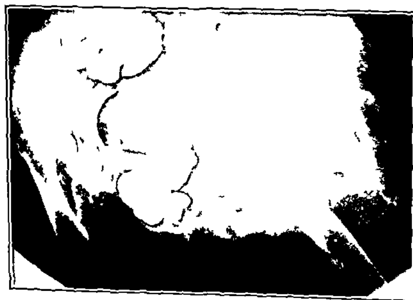


Fig. 247. Neurogenic Arthropathy. There is advanced destruction of the hip joints. Both hip joints are dislocated. The bones show advanced porosis. The patient was a paraplegic. The barium residuals are from a previous opaque enema study.

has been termed paraosteoarthropathy, ossifying fibromyopathy or myositis ossificans circumscripta neurotica The most satisfactory name is soft tissue calcification and ossification in cord lesions The mechanism of the formation of the calcification is not clearly understood Studies of the blood chemistry reveal no significant deviation from the normal in the calcium, phosphorus and phosphatase levels Numerous theories have been advanced to explain the formation of the calcific depositions In the presence of hemorrhage, there is infection and a local increase in lime salts which cause conditions favorable for the formation of ossification However, neither hemorrhage nor infection is present or precedes the bone formation in these cases The association of osteoporosis and soft tissue calcification has led to the assumption that excessive calcium mobilization predisposes to the ossification Other factors must be present since the ossifications develop only in the paralyzed parts of the body They do not occur in disuse bony atrophy, senile osteoporosis, or other types of osteoporosis The level of the injury to the cord is significant In cervical cord lesions calcifications occur in practically all cases and in dorsal cord lesions there is a very high incidence of calcification Lesions of the lower dorsal spine and the lumbar spine show calcification in a much smaller percentage

The calcifications once formed are permanent and restoration of function of the paralyzed limb does not result in resolution of the osseous depositions The involvement is extra articular The calcifications are at first amorphous, mottled and irregular in outline With the passage of time, the shadows become bony in character, present characteristic trabeculae and assume the appearance of cancellous bone The newly formed bone may be discrete or in the form of linear sheets or bands separated by irregular areas of radiance extending along the muscle and fascial planes In the region of the hip, the deposits of calcium usually are most marked above and anteriorly to the necks of the femurs and the greater trochanters Scattered masses also accumulate in the region of the tuberosities of the ischiums, about the hip joints along the femoral shafts and in the region of the knee joints particularly the collateral tibial ligaments The changes are very similar to those in ossifying hematomas The hip joint and the upper portion of the femur to the level of the lesser trochanter may be completely encased in the bone In the region of the knee the ossifications are most prominent in the medial collateral ligament and in some cases extend upward along the distal third of the shafts of the femurs They may also appear along the lateral aspect of the knee

Para-articular Calcification after Acute Anterior Poliomyelitis

Para articular calcification similar to that developing after lesions of the spinal cord and cauda equina may occur in acute anterior poliomyelitis The first case was described by Dieckmann in 1927 Freiberg reports a case in a woman thirty years of age with diffuse calcifications in the metacarpophalangeal joints the shoulders and the hips The areas of ossification did not correspond to any particular muscle or fascial plane involvement Subsequent observations revealed progressive ossification with ankylosis of the shoulders Four similar cases have been recorded in the literature *none with involvement of the hands* The para articular calcifications are considered secondary to the lesion of the central nervous system

GOUT

Gout is a metabolic disease of unknown etiology characterized by arthritis, increased concentration of uric acid in the serum and body fluids deposition of sodium urate crystals in articular, periarticular and subcutaneous tissues, bone and the urinary tract and degenerative changes particularly in the vessels of the kidneys It is believed to be due to a disturbance in the metabolism of uric acid More than 90 per cent of the cases are in males over thirty years of age It occurs in various races and nationalities and at all social levels The pattern of the disease appears to be inherited or to originate in early life The trait of hyperuricemia is believed to be inherited through a single autosomal gene which is dominant However, gouty arthritis develops in only a portion of the bearers of the gouty trait Female carriers of the gene associated with gout usually do not show an abnormally elevated plasma urate level



Fig 248 Gout There are advanced arthritic changes with destruction of the metatsarophalangeal joint of the great toe The soft tissues about this joint are markedly thickened and swollen and there are multiple irregular amorphous depositions of calcium in the periarticular soft tissues

until the menopause or shortly before and the symptoms of the disease in women seldom are manifested until about the time of the menopause In women in whom symptoms develop prior to the menopause the attacks usually begin during the menses when estrogen levels are at the lowest The uric acid level of the blood serum is above normal in 95 per cent of patients with gout The acute attacks usually begin in the third to sixth decades and are characterized by sudden onset short duration and self-limitation The attacks most commonly affect the first metatarsophalangeal joint Tophi, renal calculi and olecranon bursitis are common The late stage of gout is characterized by joint deformities which become permanent unless proper corrective measures are instituted The deformities are usually asymmetrical but may occasionally be bilaterally symmetrical and simulate rheumatoid arthritis Large subcutaneous tophi with draining sinuses and chronic nephritis are common Acute attacks of gouty arthritis in persons who are afflicted with the underlying metabolic disturbance are precipitated by trauma surgical operations systemic infections emotional stress excessive intake of nitrogenous foods or alcohol and other factors

The articular symptoms often develop suddenly The affected joints become red, warm swollen and exquisitely tender to touch and are acutely painful, especially when moved Weight bearing is impossible in the severer attacks that affect the lower extremities A moderate elevation of temperature, leukocytosis, and an increased sedimentation rate may be present The metatarsophalangeal joint of the great toe is affected in about 50 per cent of the patients, but other joints of the feet, ankles knees, hands, wrists or elbows may be involved The arthritis may be monarticular or migratory and polyarticular Joint effusion or acute bursitis not uncommonly is associated with the synovitis A first attack of acute arthritis occurring in a man past the age of thirty-five strongly suggests gout Recurring attacks of acute joint pain with completely asymptomatic intervals are infrequent in adults except in gout

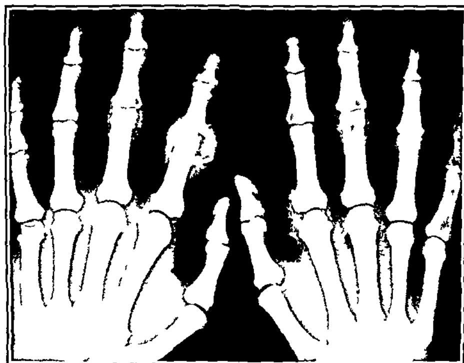


FIG 249 Gout There are extensive destructive changes about the proximal interphalangeal joints of the right index finger Charactersitic punched out areas of rarefaction are visualized in the proximal end of the middle phalanx of the right middle finger and the proximal end of the proximal phalanx of the left index finger There is marked swelling and thickening of the soft tissues of the joints of the fingers

Laboratory Findings An increase in the content of urates in the serum is characteristic of gout The level in the serum usually is above 6 mg per 100 cc Other conditions that may be associated with an increase in serum uric acid include renal insufficiency, polycythemia vera leukemia multiple myeloma pernicious anemia and certain infections Tophi containing sodium urate crystals are typical of gout They occur oftenest about the margin of the ears and in the feet and less often around the joints of the hands the olecranon and the prepatellar bursas

Roentgen Manifestations The early roentgen changes usually occur at the first metatarsophalangeal joint and consist of soft tissue swelling and an area of increased radiance at the medial aspect of the proximal end of the proximal phalanx and the distal end of the first metatarsal As the

lesion progresses there is intensification of the rarefaction with the development of a cystic or "punched out" lesion and narrowing of the joint space. The joint surfaces become markedly irregular due to destruction of the cartilage and osteophytes develop about the margins of the joint. Gouty deposits at the ends of bones cause marked expansion with destructive changes which are more extensive than those associated with deposits in the joints alone. The affected joints may be completely obliterated and fragments of bone extend into the soft tissues. Erosions, bony proliferation, and pathologic fractures may occur. The gouty deposits may cause spreading of the cortex with an appearance closely similar to that in giant cell tumor. There may be severe clinical manifestations without

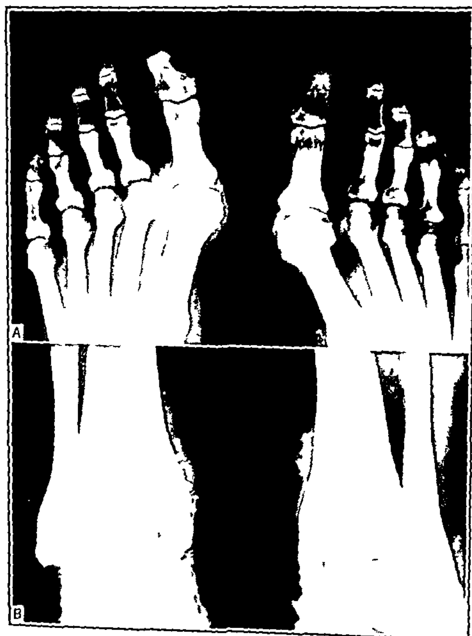


Fig. 250. Gout. *A*, Anteroposterior view of the Feet. There is swelling and thickening of the soft tissues about the metatarsophalangeal joints of the great toes with multiple calcific depositions adjacent to the joint on the left. *B*, Anteroposterior and lateral studies of the right ankle. The soft tissues overlying the anteromedial aspect of the lower end of the tibia show extensive swelling with scattered calcific depositions.

demonstrable roentgen changes In gouty arthritis, osteoporosis may develop very rapidly Tophi are not opaque to the roentgen rays and when confined to the soft tissues are manifested only as localized swellings and thickenings of the tissues The fully established cases are characterized by the presence of subchondral, endosteal, and medullary tophi The tophi show a predilection for the articular margins of the bone and there occur erosions of the articular cortex with subperiosteal outgrowths at the margins of the bone There is osteosclerosis which is usually wavy in character Late changes comprise osseous tophi walled in by compact *bone and coarse buttressing with small honeycomb translucencies* Secondary osteoarthritic changes are present and there is ankylosis of the affected joints

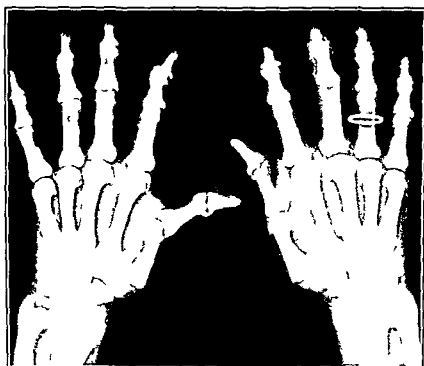


Fig 251 Osteoarthritis of the Hands There are advanced osteoarthritic changes with spur formations about the margins of the joints The interphalangeal joints are narrowed and irregular in outline There is swelling of the soft tissues about the interphalangeal joints

HYPERTROPHIC ARTHRITIS OSTEOARTHRITIS DEGENERATIVE ARTHRITIS ARTHRITIS DEFORMANS DEGENERATIVE JOINT DISEASE

Hypertrophic arthritis is also known as osteoarthritis, arthritis deformans chronic senescent arthritis and degenerative joint disease The cause is unknown It is characterized in the early stages by degenerative changes in the cartilage There is an apparent correlation between the severity of the process and advancing age the majority of people over sixty years old presenting manifestations of the disease Excessive and abnormal use of the joints are important factors in the production of hypertrophic arthritis, as are also congenital and developmental abnormalities trauma infarction previous arthritis of other types, neurogenic disturbances endocrine imbalance and other conditions True ankylosis does not occur in hypertrophic arthritis although calcification

and bony bridging may develop in the sacroiliac and acromioclavicular joints which are composed of fibrocartilage and in the interspinous ligaments with resultant limitation of mobility or rigidity The condition even in the advanced stages, may exist without causing symptoms In the interphalangeal joints of the fingers tenderness fluctuant swellings deformities, and periarticular enlargements are characteristic manifestations of the disease The knees hips, wrists, shoulders and other joints of the extremities are less commonly affected

Roentgen examination shows bony spurs, lipping or osteophyte formation about the margins of the joints The spurs are beak shaped and occur at the sites of insertion of the capsular ligaments at the joint margins at which stress is increased There is bony bridging or outgrowths in the direction of the plane of the joint Separate ossicles develop in the capsular ligaments in the interphalangeal joints Hypertrophic outgrowths of the terminal phalanges comprise the so called Heberden s nodes Osteosclerosis develops in subarticular bone with coarsening of the trabeculae along the lines of stress The late changes comprise gross loss of cartilage, eburnation with pitting of the articular surfaces subarticular cystic degenerations and subluxations

Osteoarthritis of the Spine

Osteophytes about the margins of the vertebrae thinning of the intervertebral discs arthritis of the posterior spinal articulations and postural changes may diminish the size of the intervertebral foramens Spurs along the anterior borders of the vertebrae are usually not of great clinical significance Osteophytes occurring at the posterior aspects of the vertebral bodies however, are a frequent cause of radicular pain since they are apt to encroach on the neural foramens The intervertebral discs may become narrowed with consequent diminution of the spaces between the vertebrae due to various factors As a result of trauma, the nucleus pulposus may be herniated, the disc ruptured, or the cartilage injured With advancing age or in consequence of occupational trauma the intervertebral discs lose their elasticity and become flattened Since the disc receives its blood supply from the adjacent bone, disease of the vertebral body eventually interferes with the nutrition, hence the changes once established are permanent A close correlation between thinning of the intervertebral discs and radicular symptoms has been demonstrated Oppenheimer in a series of 200 apparently healthy persons found only 22 (11 per cent) with disc narrowing On the other hand in a series of 312 unselected cases of narrowed intervertebral discs he reported clinical signs or symptoms pointing to a lesion of the involved spinal roots in 267 (85 per cent)

Osteoarthritis of the intervertebral joints, usually associated with disc thinning frequently results in narrowing of the apophyseal joints In the early stage, there is a strain on the capsule and ligaments with periarticular soft tissue reactions that may cause compression of the nerve roots Later eburnation and exostosis formation develop Joint abnormalities occur in over 20 per cent of cases with disc thinning Hadley states that changes in the posterior articulations of the spine produced pain due to nerve root compression in 72 per cent of his cases Osteoporosis occurs when the spine becomes rigid or loses its mobility The degree of decalcification may serve in a general manner as a guide to the

degree or duration of the process Scoliosis is generally associated with rotations or lists causing strain of the ligaments, intervertebral discs and articulations Disc narrowings and marginal osteophytes may occur in long standing scoliosis The cervical spine, which normally presents a moderate lordosis, may become straight less frequently more lordotic and rarely kyphotic In the dorsal region, increased kyphosis is the rule



Fig 252 Osteoarthritis of the Cervical Spine There are moderately advanced osteoarthritic changes with bony spur formations about the margins of the bodies of the fifth sixth and seventh cervical vertebræ The spurs are present along the anterior and the posterior margins of the vertebral bodies The cervical spine is straight with practically complete absence of the normal anterior curvature



Fig 253 Osteoarthritis of the Lumbar Spine There are hypertrophic changes with spur formations about the margins of the bodies of the vertebræ

The literature on radiculitis due to disease or injury of the vertebrae and protrusions of the intervertebral discs is vast, it is meager, however on the subject of hypertrophic arthritis and postural strain In consequence the practitioner has little opportunity to become familiar with the clinical manifestations of this condition, and it is not generally appreciated how commonly pain in the anterior portion of the chest may be of spinal origin This pain may occur in the form of severe attacks closely simulating coronary artery disease Coronary artery disease and hypertrophic arthritis of the spine with radicular symptoms may coexist in the same patient for both are common with advancing age It is known that hypertrophic arthritis of the spine may be present without producing clinical manifestations The roentgenologic demonstration of osteoarthritis may be of great aid in establishing the diagnosis of radiculitis This is particularly true when the symptoms are of short duration, the attacks cannot be reproduced, or the radicular characteristics of the pain are not clear cut

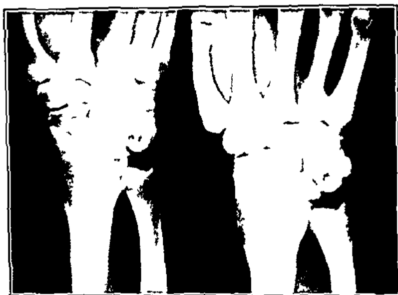


FIG 254 Cystic Changes in the Carpal Bones

Osseous Cyst-like Lesions

Cyst like areas of rarefaction are common in the bones about the hip and other joints in patients with degenerative arthritis and in the radio carpal regions The cysts vary widely in size, shape and location There is usually no sclerosis of the surrounding bone The adjacent articular cartilages may be narrowed and the bone cortex obscured The cystic lesions may occur in association with fractures of the carpal and meta carpal bones The changes which develop in bone after interference with the blood supply are those of progressive and in some instances complete replacement by fibrous tissue the areas of necrosis being resorbed without subsequent replacement by living bone These areas form the cysts seen in the late stages of the disease The degenerated articular cartilage is replaced by fibrocartilage and fibrous tissue On histopathologic examination, the cyst like areas in the bones about the wrist are similar to those in the upper ends of the femur and the pathogenesis is essentially similar

Bugnion investigated a series of 300 subjects postmortem and found lacunar formations in the bones of the wrist in almost half of the cases

The cysts were most frequent in the capitate and lunate. Sex and occupation did not appear significant. Age is an important factor, the incidence of the changes increasing with advancing years and reaching a maximum in the sixth and seventh decades. He describes three forms of carpal lesions: (1) cysts which develop as a result of herniation of the synovial membrane into the bone usually measure 2 to 3 mm. in diameter and form in sites of lowered resistance, usually at the insertions of the ligaments and points of penetration of blood vessels; (2) necrotic cysts due to vascular disturbances may measure 1 cm. or more in diameter and occur in the aged; (3) the arthritic marginal cysts may comprise a cartilaginous lesion secondary to cystic formations beneath the articular cartilages or take the form of a marginal laminar osteochondrosis due to ischemic disturbances. The various types may occur simultaneously.

INTERMITTENT HYDRARTHROSIS

Intermittent hydrarthrosis is a chronic disorder of recurring effusions usually involving the knees and accompanied by minimal local or constitutional symptoms. The effusions are periodic in pattern, usually occurring every seven to eleven days. The cause is unknown. In some cases the effusions are prodromal to typical rheumatoid arthritis, while other cases have been reported in which the typical recurrent pattern was present for as long as twenty-two years. The condition is uncommon and the diagnosis should be made with caution after observation and careful study to rule out other more frequent causes of effusion.

HEMOPHILIC ARTHRITIS

Hemophilia is of great importance to the radiologist because of the occurrence of joint involvement which very closely simulates other diseases. It is a disorder characterized by prolonged and immoderate hemorrhage dependent upon delayed coagulation of the blood. The condition occurs only in the male but is transmitted through the female. The disease occurs in two stages. The first stage is characterized by an acute hemarthrosis. The hemorrhage may be spontaneous or traumatic in origin. The bones may be involved in association with the arthritic manifestations or independently. The affected joint becomes markedly distended with blood. The second stage is that of chronic degenerative arthritis and ensues after one or more hemarthroses. In many cases there are no demonstrable roentgen changes. The knees and the elbows are most frequently affected. The extent of involvement increases in proportion with the age of the patient. In young individuals there is little or no joint involvement. Patients in the thirty to fifty year age groups show involvement of the joints in practically all cases.

There are two factors of importance with reference to the changes in the joints, mechanical and chemical. The accumulation of blood within the joint cavity causes a marked cellular reaction in the synovial membrane. The macrophages phagocytose the red cells and accumulate in the subsynovial tissues beneath the capsule. There is a tendency to fibrous ankylosis due to thickening and contracture of the joint capsule. Degenerated iron pigment deposits may cast dense mottled roentgen shadows and are easily mistaken for calcifications. The cartilage in

chronic hemophilic arthritis becomes irregularly serrated due to localized absorption and subsequent proliferation The joint spaces become narrowed, often at points other than those of maximum weight bearing The periphery of the articular cartilage shows spurring due to the formation of osteophytes The characteristic changes in the bones are those of subarticular cystic absorption Diffuse decalcification of the bone because of disuse accentuates the cystic changes The size of the cysts varies from less than a millimeter to over a centimeter in diameter The cysts

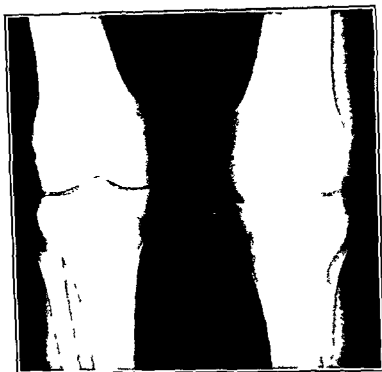


FIG 255



FIG 256

FIG 255 Hemophilic Arthritis of the Knee The articular surfaces of the lower end of the left femur are markedly irregular in outline and there is an area of rarefaction in the intercondylar region The soft tissues about the left knee are thickened and swollen The patient was a hemophiliac with hemorrhage into the knee joint

FIG 256 Hemophilic Arthritis of Knee The knee joint space is markedly narrowed There is irregularity of outline and eburnation of the articular surfaces of the femur and tibia with osteoporosis of the bones of the knee The patient was a hemophiliac with a history of repeated hemorrhages into the joints

may be multiple and irregular in outline or loculated Hemophilic blood exerts an unknown chemical effect on bone tissue The intraosseous hemorrhage causes an aseptic necrosis of bone In many instances there is a history of previous hemorrhage(s) into the joints with no roentgen manifestations of abnormalities in these joints

Roentgen Manifestations Fluid in the joint is the first manifestation and is characterized by haziness and slightly increased density in and about the joint Gradual erosion of articular cartilage occurs with resultant narrowing and irregularity of the joint space Rounded areas of bone destruction appear beneath the joint cartilage and the cartilage may be

completely destroyed. The capsule becomes markedly thickened, probably due to the deposition of hemosiderin in and about the joint. Spurs form at the margins of the joints and dislocations may occur. Cystic areas of increased radiance in the bones adjacent to the joints are the most characteristic manifestations of hemophilia. Areas of calcific density about the joints may be the result of ossification of the hemorrhages or precipitation of the hemosiderin in the clotted blood. Cavities may develop in the bones at points removed from the joints also due to the presence of blood and fibrous tissue. The intercondylar fossa becomes deepened and widened in the case of involvement of the knee. Bony ankylosis is rare. Transverse bands of increased density may occur in the metaphyses. The knee is most commonly affected followed in order by the elbow, ankle, wrist, shoulder and hip. Multiple joint involvement is common, and lesions may occur in three, four, or more articulations. The joint changes may develop during the first or second year of life when the child is first beginning to walk. The most common time of appearance, however, is at puberty.

Ghormley and Kent describe changes in the shafts of the long bones in a series of cases. While these manifestations are much less common than the joint changes they may be more severe. There may be involvement of the phalanges, the olecranon and the femur. In all instances the involvement affects the adjacent joint to some extent. It is not certain whether the bone changes are secondary to hemorrhages into the joint or are due to primary hemorrhages in the bone or the subperiosteal tissue. The case with involvement of the phalanges of the thumb showed a large tumor-like shadow with destruction of the terminal phalanx. In the case involving the femur there were varying amounts of destruction of the cortex and in the case involving the olecranon a cyst-like area was present in the olecranon process. The pseudotumor may arise from (1) hemorrhages originating in the joint and extending along the bone to produce erosion, (2) subperiosteal hemorrhages which lead to formation of new bone and subsequently to absorption and destruction of bone, and (3) cortical or medullary hemorrhage which causes cystic changes, later destroys the bone and results in further hemorrhage. It is difficult to state accurately in any individual case the exact sequence of events. It is important to recognize the condition early so that destructive changes may be reduced to a minimum and controlled.

Peterson reports a case of hemophilia which showed characteristic changes in the knee joints and in addition unusual osseous changes with a large cyst-like rarefaction in the squama of the left iliac bone. The defect measured 9 × 11 cm. in diameter and the medial margin was sharply defined and slightly sclerotic. Laterally the demarcation was indefinite and there was a rounded expansion of the bone. Within the area of rarefaction there was a faint trabecular pattern. The hip joints revealed no abnormality. In the space between the neck of the femur and the squama of the left iliac bone there was a faint irregular opacity, presumably an indication of beginning calcareous deposit. Extending laterally downwards from the left ischial tuberosity toward the lesser trochanter and branching in the form of a Y around the latter was a sharply defined calcareous shadow about 1 cm. in width. Along the medial aspect of the right hip joint a similar shadow extended from the upper part of the joint toward the lesser trochanter. Re-examination subsequently showed the cyst-like rarefaction more sharply defined and the

margins more sclerotic The lateral margin was slightly more irregular and appeared expanded At this time there was also found an area of rarefaction about 2 cm in diameter immediately below the upper margin of the iliac crest The area of calcification between the left femur and the squama of the iliac bone was more marked The other calcific depositions were unchanged The bony changes were due to subperiosteal intra osseous hemorrhage with subsequent cystic degeneration

Differential Diagnosis The conditions which must be considered in differential diagnosis are acute hemarthrosis and chronic degenerative arthritis In acute hemophilic hemarthrosis the changes are similar to those in other types of serous or purulent synovitis in which the joint space has become distended with fluid The joint is held in partial flexion since the capsule is under lessened tension in this position, There is generalized thickening and increased density of the periarticular tissues the changes being most marked in the region of the capsule At this stage roentgen diagnosis is not possible as the manifestations do not differ from the hemarthroses which occur subsequent to trauma in normal individuals or spontaneously in hemorrhagic disease In the chronic stage, accurate differential diagnosis is more frequently possible Osteoarthritis usually occurs after the age of forty five while in hemophilia the roentgen changes are frequently present in patients who are much younger Cystic changes in the bones may occur in both conditions They are the rule in hemophilia but are less common in osteoarthritis Generalized decalcification is more frequent and usually is more marked in hemophilia than in osteo arthritis Differentiation from rheumatoid arthritis may be difficult In rheumatoid arthritis the wrists and the phalanges are more frequently involved and bony ankylosis of the joints is common Subchondral cystic changes are much more frequent in hemophilia than in rheumatoid arthritis Tuberculous arthritis is a more difficult problem in differential diagnosis In tuberculosis, the articular surfaces do not show fine etching ragged irregular scalloping being the rule It is the duty of the roentgenologist to stress the possibility of hemophilic arthritis to the clinician in order that the diagnosis may be established if surgery is being considered The diagnosis of hemophilia is dependent on the demonstration of a prolonged venous blood coagulation time However, in a young male with a family history of a bleeding tendency and joint involvement, a highly presumptive diagnosis may be made The roentgen changes are an important aid in diagnosis

ADDITIONAL READING

GHORMLEY R K and CLEGG R S Bone and Joint Changes in Hemophilia with Report of Cases of So called Hemophilic Pseudotumor J Bone & Jt Surg *30A* 589 1948

McDONALD F J and LOZNER E L Hemophilic Arthritis A J Roent *49* 405 1943

PETERSON J A Case of Osseous Changes in a Patient with Hemophilia Acta Radiol *28* 323 1947

ALKAPTONURIC ARTHRITIS OCHRONOTIC ARTHRITIS

Ochronotic arthritis is a rare metabolic disturbance manifested by a diagnostic triad (1) blue-black cartilages, especially of the ears and nose brownish discoloration of the scleras and gray blue pigmentation of the skin (2) urine which turns dark brown or black on standing or alkalinization and (3) arthritis The first case was described in postmortem

findings by Virchow in 1866 In 1904 Osler reported the first living case Alkaptonuria is characterized by the appearance of alkapton or homogentisic acid in the urine It is considered an inborn error of metabolism of certain amino acids, namely tyrosin and phenylalanine About 160 cases have been reported in the literature and the approximate incidence is about 1 in 10 million of the population Apart from the changes in the urine the condition does not usually give rise to symptoms of clinical significance However, in about 50 per cent of the cases the urinary changes have been accompanied by ochronosis and degenerative arthritis The first accurate description of this condition is attributed to Boedeker in 1859 He discovered in the urine of a patient with glycosuria a second reducing substance which he termed alkapton because the urine turned black when alkalies were added to it In 1891 Wolkow and Baumann isolated this substance and identified it as homogentisic acid The condition has been classified with other inborn errors of metabolism such as albinism cystinuria and pentosuria It is characterized by the presence of homogentisic acid in the urine and is inherited as a mendelian recessive character due to a defect in a single gene It is a prime example of a metabolic defect which behaves in accordance with the mendelian law The condition is more frequent in males than in females in the proportion of about 2 to 1 In 42 per cent of the reported cases, there has been a history of consanguinous marriage Alkaptonuria is present from birth and persists throughout life It may be suspected from infancy because of the blackening of the urine and staining of diapers or clothing although frequently it is unrecognized until later in life The condition is frequently incorrectly diagnosed as diabetes because solutions containing copper as in Benedict s or Fehling s solution are reduced An important point is that the acid is excreted by the sweat glands and clothing coming into contact with the axillary perspiration is stained black The blood Wassermann reaction is positive in some instances

Ochronosis is characterized by the presence of pigmentation of certain bodies or granules which appear ochre in color under the microscope hence the name ochronosis Albrecht in 1902 was the first to suggest that alkaptonuria was the cause of ochronosis Most cases of ochronosis occurring with alkaptonuria have been in patients over forty The pigment occurs in the cartilage tendon fatty tissue endocranium intima of large vessels kidney tonsil lung and thyroid There may be a butterfly shaped area of pigmentation The cartilages are pigmented and may present a grey discoloration An early sign is the appearance of brown patches in the sclera These are usually triangular in shape with their bases toward the cornea The nasal cartilages may be stained blue and there may be a blue tint in the knuckles and hypothenar eminences in the late stages The accumulation of pigment is very gradual and it may be several years before it becomes prominent enough to be recognized Conditions other than alkaptonuria may produce ochronosis Melanin in the urine or melanuria may occur after the external use of phenol This is termed carboluria The association of osteoarthritic lesions with alkaptonuria is important in diagnosis The condition frequently occurs in several members of the same family Garrod states that ochronosis of the articular cartilage is not the cause of arthritis in every instance as the cartilages of the affected joints in some cases show slight if any staining Also intense blackening of the articular cartilages may occur without other joint changes

Pathology The pathological process in the affected joints is described by Hertzberg as follows The ochronotic pigment is deposited in the articular cartilage, especially in old cartilage with poor metabolism This becomes black, loses elasticity turns brittle, and shows little resistance to mechanical strain The cartilage cracks easily and small black fragments may be detached into the joint where they soon become imbedded and attached to the synovial membrane They are usually firmly fixed to the synovia Mechanical strain leads to changes in the articular surfaces with sclerosis and the formation of cysts and small infractions The subchondral bone marrow is irritated, proliferates, and grows into the diseased cartilage Marginal exostoses are formed Pigmented cartilage is reported to be able to become forced into the marrow cavity, being sur-



Fig 257 Alkaptonuric Arthritis *A* Right shoulder *B* Left knee *C* Lumbosacral spine pelvis and hips *D* Lumbar spine lateral projection There is advanced degenerative arthritis involving all the joints The intervertebral spaces are markedly narrowed There are wafer like calcifications of the intervertebral discs with marginal osteophytes about the vertebræ

rounded there by growing granulation tissue The cartilage may disappear, leading to bony ankylosis Osteoporosis may be present The pigment is deposited much less in the bony system than in the cartilage Lacunar resorption occurs and these patients are frequently subject to fractures as a result of minor injuries The morbid processes greatly resemble those observed in ordinary deforming osteoarthritis On gross and histopathologic study, the cartilages, ligaments tendons, intima and in some instances the bones are grossly black and show ochre microscopically (Virchow) It is the deposition of these substances in the articular cartilages which hastens premature degeneration of the joints The endocardium may be discolored and pigmented concretions are present in the mitral and aortic valves "

Clinical Manifestations Clinically, the patients have a peculiar stance and gait due to the involvement of the joints of the lower limbs and spine In the spine, there is rigidity and kyphosis as in spondylitis deformans There are alternating areas of osteoporosis and osteosclerosis The patients assume a posture which suggests Paget's disease The intervertebral discs are dense due to secondary calcifications following degeneration of the cartilage resulting from the deposition of homogentisic acid There is a metabolic injury to the articular cartilage which results in gross degenerative osteoarthritis The arthritis rarely develops prior to the age of forty and at this time the process is well advanced Many of the patients present themselves with early symptoms which suggest the presence of a prolapsed disc The gross destructive changes in a non-weight bearing joint such as the shoulder closely resemble those in neuropathic arthritis

Roentgen Findings The roentgen examination may show evidence of extensive degenerative osteoarthritis in the hips, knees shoulders elbows, spine and other joints The characteristic manifestation is the wafer like calcification of the intervertebral disc in association with marginal osteophytes There is involvement throughout the entire spine from the cervical to the lower lumbar region The condition has best been characterized by Hench as resembling rheumatoid arthritis clinically and osteoarthritis radiographically Other affected joints present typical osteoarthritic changes with a tendency toward osteoporosis, large spur formations and marked narrowing of the joint spaces The arthritis is an integral part of the complex not an occasional manifestation It is chronic progressive shows frequent exacerbations, and is accompanied by synovial effusions The sacroiliac joints are sclerotic Rounded areas of increased radiance in the pelvic bones upper humeri and condyles of the femur are due to ochronotic deposits

Treatment Therapy consists of parenteral administration of liver extract, ascorbic acid and tocopherol Salicylates and aminopyrine have also been utilized The results of these measures have been inconclusive Cortisone has afforded temporary relief the joint manifestations and reducing substances in the urine reappearing after cessation of the treatment

PSORIATIC ARTHRITIS

There is a type of arthritis which is peculiar to persons afflicted with psoriasis and occurs particularly in patients with the pustular form of the disease The condition has a predilection for the distal joints of the hands

and feet, although other joints may be affected also Psoriasis occurs in about 3 per cent of patients with typical rheumatoid arthritis This probably indicates a tendency for the two diseases to coexist There are certain instances of psoriasis in which arthritic changes appear in the terminal interphalangeal joints of the fingers showing psoriatic nail lesions These may represent a distinct form of arthritis The roentgen manifestations are characteristic and differ from those of rheumatoid arthritis The earliest changes occur in the interphalangeal articulations and comprise marginal erosions at the edges of the phalanges Irregular areas of destruction develop and extend along the shafts of the bones imparting a scalloped appearance to the cortex with numerous punched out defects Later in the course of the disease there is gross destruction of the terminal aspects of the phalanges The ends of the bones may in the advanced case become extremely thin and taper to a point There is

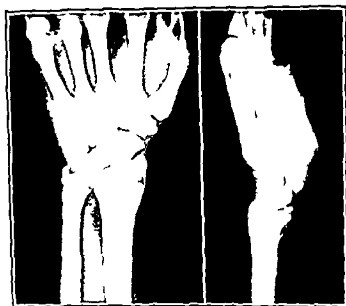


Fig 258 Arthritis in a Patient with Psoriasis The carpal joint spaces are narrowed hazy and irregular in outline The soft tissues about the wrist and the proximal portion of the hand are markedly thickened and swollen The patient had advanced psoriasis of twenty years standing with associated arthritis

usually little or no evidence of healing In some instances there is overgrowth at the tendon insertions with the production of cup like deformities Irregular nodules of new bone formation may form adjacent to the affected joints In the very late stages of the disease and much later than in rheumatoid arthritis ankylosis develops in the joints affected by the disease The metacarpophalangeal joints are usually spared The radioulnar joints are frequently involved and may be subluxated The metatarsophalangeal joints also may be dislocated, the phalanx undergoing proximal and upward displacement

REITER S DISEASE

Reiter s disease is a syndrome of urethritis conjunctivitis and arthritis and must be differentiated from gonococcic infection It occurs only in males There is unsubstantiated evidence that the causative agent is a

pleuro pneumonia like *organism Any of the three systems involved* may present the major symptoms and progress to major sequelae Usually the urethritis is of a slight mucopurulent type and the conjunctivitis of a catarrhal type, with the joint disorder predominating Urethral smears and cultures are usually negative for specific microorganisms The disease is self limited but tends to recur Permanent damage to the joints is rare

MANDIBULAR JOINT NEURALGIA

(Costen s Syndrome)

Neuralgia of the temporomandibular joint was first reported by Costen in 1934 and has become established as a definite clinical entity It is an important clinical syndrome and must be considered in the differential diagnosis of every case of recurrent facial pain The condition may be due to malocclusion from any one of a variety of causes or destructive changes involving the temporomandibular joint and is characterized by vertex occipital, and facial pain, otalgia glossodynia and disturbance of function of the affected joint There is abnormal pressure in the man dibular fossa with in many instances partial or complete closure of the internal auditory canal, direct nerve compression within the abnormal *joint reflex irritation of the nerves which lie in close relation to the joint* and muscle spasm The recognition of the syndrome is usually dependent on its being considered in the differential diagnosis in all obscure cases of pain in the region of the jaw ear eye nose, and face Patients with bizarre facial and head pain are considered as a rule to have sinusitis trigeminal neuralgia dental disease and other lesions Many are dubbed psychoneurotics The recognition of Costen s syndrome will eliminate much suffering and shorten the patient s illness as correction of the cause of malfunction of the temporomandibular joint results in cure of the condition The commonest etiologic factor is malocclusion of the teeth Other frequent causes are ill fitting dentures destructive lesions of the *temporomandibular joint impacted third molar teeth and similar local* pathology The clinical manifestations in addition to the pain are numerous and protean There may be impaired hearing tinnitus dizzi ness pain in the ear occiput vertex *pharynx nose and glossodynia* Eating laughing yawning and other activities which result in stress of the temporomandibular joint aggravate the symptoms On physical examination there may be found overclosure of the jaws crepitus within the temporomandibular joint tenderness in the region of the joint un even jerky movements on opening and closing the jaw and trismus

Roentgen examination reveals the cause of the malocclusion and local changes in the temporomandibular joints and the teeth The joint may show *alterations of density narrowing or widening of the joint space* irregularity of outline and fixation of one condyle against the tubercle Deepening of the fossa is a manifestation of abnormal stress The roent genograms must be made both with the mouth open and closed and both sides must be studied for comparison as one side may function normally while the other shows abnormalities of position contour or degree of mobility

DISEASES OF THE PERIARTICULAR REGIONS

HYPERTROPHIC OSTEOARTHROPATHY

Hypertrophic osteoarthropathy occurs in two forms The more common variety of the disease is found in association with primary diseases of the lung, heart, liver, blood, and mediastinum and is therefore termed secondary hypertrophic osteoarthropathy A less frequent form, characterized by a familial tendency and absence of underlying or associated disease elsewhere in the body is termed primary or idiopathic hypertrophic osteoarthropathy While both types have many features in common there are certain fundamental differences which necessitate careful distinction

SECONDARY HYPERTROPHIC OSTEOARTHROPATHY

The first report of this condition was in 1889 by Marie and Bamgerber It is now generally appreciated that it does not always result from primary disease of the lung and may also occur in diseases of the heart, liver and blood and carcinoma of the thymus Similar changes have been reported in the rat dog horse and lion Most of the cases in dogs were secondary to pulmonary tuberculosis and bronchiogenic carcinoma The lesion is frequently confused with disease of the joints In many instances there are no symptoms referable to the chest Discovery of the changes in the bones is an indication for careful study of the chest and frequently discloses a previously unsuspected pulmonary neoplasm or other disease of the chest There is frequently associated clubbing of the fingers The clubbing and hypertrophic osteoarthropathy arise from the same causes the latter representing an advanced stage of the clubbing Lung abscess, empyema tuberculosis and bronchiectasis were formerly believed to be the predominating causes of hypertrophic osteoarthropathy Recent reports show that carcinoma of the lung is a more prevalent and important cause If all patients with carcinoma of the lung have roentgen study of the extremities the incidence of hypertrophic osteoarthropathy will doubtless be much higher than is generally believed since this condition may be present for a long period without producing clinical manifestations The presence of joint symptoms in previously healthy individuals should be carefully evaluated in order to determine the possibility of coexisting underlying pulmonary disease In many instances, the associated disease is first manifested by painful or swollen joints and roentgen examination of the joints often leads to the discovery of periosteal new bone formation subsequent investigation establishing the proper diagnosis

Pathology On gross examination the changes are essentially those of chronic proliferative subperiosteal osteitis surrounding the shafts of the bones The lesion is most common in the tibia the fibula the radius the ulna and less frequent in the humerus, femur and phalanges In advanced cases the clavicles, spines of the scapulas and vertebrae may be involved The ends of the shafts usually are not affected The new bone formation varies from 1 to 7 mm in thickness The original cortex remains intact The new bone formation is extremely vascular The clubbing of the fingers is due to soft tissue changes and frequently exists without bone involvement In rare instances, absorption of the terminal

phalanges may occur, being preceded by hypertrophic changes There may be practically complete absorption of the ungual process and the terminal phalanges assume the appearance of a thumb tack The absorption may progress to such a marked degree that only the base of the terminal phalanx remains The joints are involved in a large percentage of the cases The joint changes are those of synovitis, erosion, and ankylosis

Theory of Etiology There are many theories to explain this condition Experimental attempts to produce hypertrophic osteoarthropathy have proven universally unsuccessful It has been stated that chronic increase in the blood flow and increased periosteal nutrition are responsible for the proliferation of subperiosteal new bone This has never been proven experimentally Other theories include toxic absorption of infectious material and alteration of the acid-base equilibrium Fried found 4 cases of hypertrophic osteoarthropathy in patients with bronchogenic carcinoma, each of whom showed pituitary dysfunction There is no clinical evidence of obstruction to the venous return of blood Prompt and complete disappearance of the condition occurs after pneumonectomy and has also been reported after the removal of a benign intrathoracic neurofibroma At least two factors appear necessary to produce secondary osteoarthropathy, a toxemia due to long standing disease and circulatory disturbance resulting from cardiac or pulmonary involvement Since similar changes occur in persons living at high altitudes the condition may be due to lack of oxygen The disease may occur in chronic laryngeal obstruction or in association with primary occlusion of the hepatic veins

Roentgen Manifestations In early cases, clubbing of the fingers with increase in the size and density of the soft tissues of the tips of the fingers is the first manifestation At this stage the bones show no demonstrable changes Later there is expansion of the terminal phalanges with thickening of the periosteum of the shafts of the phalanges The long bones of the forearms and lower legs show irregular periosteal thickening with formation of new bone along the shafts There is marked irregularity of outline of the involved bones The medullary cavity shows no change There may be definite bone atrophy In the later stages the disease is characterized by a slowly progressive ossifying periostitis which begins in the distal end of the diaphysis and gradually extends until the entire shaft is affected The changes have been found in practically all of the bones but are most common in the long bones The distal thirds of the bones of the legs and the forearms are most apt to be involved The proliferation is greatest in the region of the diaphysis decreasing toward the metaphysis and being relatively slight in the region of the epiphysis The ungual tufts may undergo atrophy but normal variations are common The endosteum is not involved and the medullary cavity remains normal in width and contour In the late stages there may be joint involvement the joints adjacent to the affected bones usually showing arthritic changes While all of the bones are affected the change is best visualized in the long bones It appears earliest and becomes most marked in the distal third of the leg and forearm The distal phalanges are unusual in that proliferation of bone is absent or minimal and when it does occur involves only the anterior surfaces The ungual tufts as a rule undergo absorption or atrophy There is usually no involvement of the carpus, tarsus vertebra or skull The endosteum does not participate in the hypertrophy and the medullary cavity is not constricted Rarely the bone becomes thickened but endosteal absorption keeps pace with the periosteal proliferation so that the cortical width remains normal

IDIOPATHIC HYPERTROPHIC OSTEOARTHROPATHY

Chronic idiopathic or primary hypertrophic osteoarthropathy is a condition which occurs predominantly in males at the age of puberty or adolescence. It is characterized by clubbing of the digits, enlargements of the bones and joints, and thickening of the skin of the face, usually in the absence of demonstrable primary disease. The disease progresses

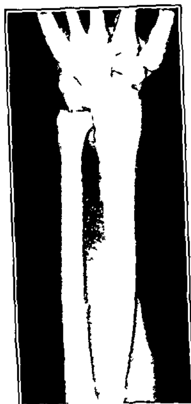


Fig 259

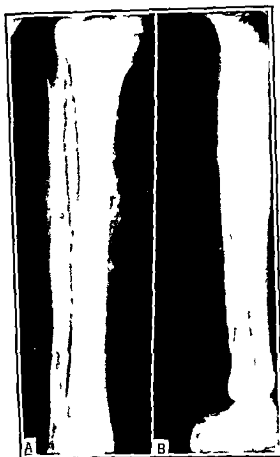


Fig 260

Fig 259 Pulmonary Osteoarthropathy There is irregularity of outline and new bone formation along the margins of the bones of the forearm and the proximal portions of the first and fifth metacarpals The patient had carcinoma of the lung

Fig 260 Primary Osteoarthropathy of Lower Leg A Anteroposterior view B Lateral view The bones of the lower leg show cortical thickening and irregularity of outline The medullary cavity of the tibia and fibula is almost completely obliterated in its middle third The patient had pachydermia There was no pulmonary disease

insidiously and deformity and marked disability ensue. The disease is familial in tendency, has a superficial resemblance to acromegaly and other forms of osteoarthropathy, and is associated with varying prominences of the skin or skeletal changes. The cause is unknown. The incidence of the disease is low. The idiopathic form comprises 3 to 5 per cent of all cases of osteoarthropathy and is important despite its rarity because it represents a pure form of osteoarthropathy not associated with the effects of primary disease elsewhere in the body.

Symptoms and Signs The primary form of the disease is characterized by onset at puberty, definite predilection for males, familial tendency and seasonal fluctuation The symptoms are characteristic The onset is gradual and insidious The course is slowly progressive with deceleration to a virtual standstill after a period of years In some instances, the changes are most prominent in the skin while in others the manifestations are most pronounced in the bones or joints Patients in whom the joints are affected show the greatest disability, while those in whom the other changes are most prominent show the greatest deformity There are few or no general manifestations There may be loss of energy with weakness and excessive sweating Secondary sexual disturbances such as hypertrophy of the male breast, feminine distribution of hair and scanty growth of beard are common Libido and potency are unimpaired In the female, there is delayed menarche Acne and seborrhea occur as a rule Fever is present only with acute episodes of joint involvement

The bone enlargements may cause symptoms There is deformity with a subjective sensation of heaviness Fractures heal and osteoporosis of disuse occurs as normally When the joints are involved there is marked enlargement, evening stiffness and vague arthralgia The enlarged joints may give rise to arthritis Effusions are common and are sterile There is marked limitation of motion of the joints The changes in the skin are striking and frequently suggest leprosy because of the thickened features and large folds which resemble the gyri of the brain The changes in the eyelids may become so marked as to require plastic procedures Similar changes have been described in mentally deficient patients in acromegaly and in certain families without other demonstrable abnormalities

Changes in the gastrointestinal tract have also been found in idiopathic pulmonary osteoarthropathy The mucosa of the stomach is coarse thickened and forms irregular dense folds which extend through the entire length of the organ Filling defects may occur in the media and antral portion of the stomach Peristalsis is sluggish or absent Gastric emptying is not affected Histopathologic studies reveal changes similar to those in chronic gastritis

Idiopathic hypertrophic osteoarthropathy has been termed familial acromegaloid osteosis idiopathic familial osteophytosis hypertrophic osteoarthropathy without other underlying disease and achropachydermia with pachyperiostitis These terms were used in the past because of certain characteristics of the disease such as its familial tendency its superficial resemblance to acromegaly its similarity to other forms of hypertrophic pulmonary osteoarthropathy and also because of the skin and skeletal changes Hereditary predisposition is a possible etiologic factor as evidenced by the alleged familial involvement in a majority of the cases previously recorded

Roentgen examination reveals enlargement of the bones without elongation especially in the fingers the lower third of the forearm and the lower two thirds of the leg There is hyperplasia of the periosteum particularly of the radius and ulna The medullary cavity is narrowed or obliterated It differs from pulmonary osteoarthropathy in that the metacarpals and the metatarsals are rarely affected The manifestations may result in confusion with acromegaly and the lesion has been termed pseudo acromegaly There are no changes in the sella

Pathology The fundamental change appears to be a circumferential enlargement of the bone with hyperplasia of the connective tissue and fat in all the layers increased vascularity and increase in the intercellular fluid The joints show hypertrophy of the articular surfaces The bone is normal on histologic and chemical examination except for great thickening of the limiting membrane of the periosteum and the presence of a very active bone forming layer There is massive medial hypertrophy of the small peripheral arteries, the change being similar to that which occurs in hypertension Laboratory studies while not of aid in establishing the diagnosis are important to rule out other lesions The blood chemical content is normal



FIG 261 Idiopathic Hypertrophic Osteoarthropathy Acropachydermia and Pachyperiostitis *A* Lower Leg The shaft of the tibia is increased in thickness and density and presents multiple irregularities of outline *B* The Stomach The gastric outline is markedly irregular There is extensive mottling within the lumen of the stomach and the gastric mucosa is coarse and thickened

Differential Diagnosis On roentgen examination the changes in the primary and secondary forms of the disease are essentially similar The diagnosis of secondary osteoarthropathy is excluded by considering the history the familial involvement the age at onset, the duration and the characteristic progression of the changes Other diseases which may cause confusion comprise acromegaly, rheumatoid arthritis, specific and non specific periosteitis osteitis deformans and osteoporosis Secondary osteoarthropathy is the most important condition to consider in differential diagnosis since it may indicate the presence of serious underlying disease This is particularly true in relation to cancer of the lung as the initial symptoms are often those of the osteoarthropathic syndrome and many cases of bronchiogenic carcinoma first present themselves as hyper-

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the mass is highly malignant or potentially so The condition is known in the literature under many names the chief of which are synovial sarcoma, synovioma synovioloma and synovial fibrosarcoma The condition occurs as a single, sharply circumscribed mass which almost invariably lies in intimate relationship to one of the joints In many instances the tumor is firmly attached to the adjacent tissues at one or more points The mass may be firm and moderately fibrous or soft, spongy, and friable In some instances it is lobulated or cystic and contains a clear fluid There may be calcification within the mass Rarely the entire tumor lies within the cavity of the joint On histopathologic examination, the neoplasm shows a highly variable and complex mixture of two cellular components one of which is villous and is derived from the synovial lining while the second is connective tissue which forms a sarcomatous pattern



Fig 262 Synovioma of the Knee There is a large rounded soft tissue density in the popliteal region a synovial (Baker s) cyst Because of the irregularity of outline of the lower end of the posterior aspect of the femur the possibility of an aneurysm was considered At operation a synovioma was found

Clinical Manifestations The ages of the patients in the reported cases have ranged from ten to seventy years, the greater majority occurring between the ages of fifteen and forty five The most common site is about the knee joint Other frequent locations are the foot wrist hand hip shoulder, ankle and elbow The tumor is manifested by pain, swelling and disability There is usually a mass which is limited in its mobility In the past treatment has comprised such procedures as excision, amputation disarticulation and combined surgery and irradiation In some instances the tumor is highly malignant and rapidly growing with death ensuing in less than a year In other cases it remains stationary for many years Recurrence after therapy is frequent Metastases or direct extension to the neighboring tissues may occur early in the course of the

trophic osteoarthropathy The bone symptoms may precede pulmonary manifestations by six to twelve months and in rare instances the bone manifestations may exist for as long as two years prior to the onset of lung symptoms In secondary osteoarthropathy the bone changes may act as the barometer of the underlying disease, waxing and waning with the activity of the pulmonary lesion There may be complete disappearance of the clubbing and bony changes after spontaneous cure or surgical therapy of the underlying disease Complete remission has never been described in the hereditary or idiopathic type of the disease

Acromegaly is the antithesis of chronic osteoarthropathy as the short, tubular bones are increased in length with proportionately slender diaphyses and hypertrophied cancellous extremities The ungual tufts and the sites of tendon and ligamentous insertions are hypertrophied In rheumatoid arthritis, the early changes which occur before periosteal proliferation becomes manifested may be confusing Periosteal proliferation of the distal ends of the long bones is frequently seen during roentgen study of the wrist or knee and should establish the diagnosis Periostitis, either specific or nonspecific, may cause confusion if only a single extremity is studied Syphilitic periostitis does not show symmetrical distribution and tends to involve the tibia but not the fibula and only very rarely the bones of the forearm It is usually not concentric Nonspecific periostitis such as occurs with infections of the foot in diabetes is characterized by soft tissue change, destruction of bone in association with periosteal proliferation, endosteal proliferation and narrowing of the medullary cavity Osteitis deformans may be similar in many respects to advanced idiopathic osteoarthropathy The age of onset the presence of cystic changes or bowing of the bones the distributions of the involved bones, and the disordered trabeculation with involvement of the medullary canal usually present a typical picture In osteopetrosis there is narrowing of the medullary canal, which usually establishes the diagnosis The hereditary form of osteoarthropathy may be difficult to differentiate in patients after puberty Periosteal proliferation is present in the case of the distal long tubular bones but is never of extreme degree The history of a hereditary trait may be necessary to differentiate the primary from the early secondary type If the underlying disease is chronic slowly progressing and of long duration it may produce changes identical with those in the chronic idiopathic form of the disease The evidence of primary disease elsewhere in the body can usually be established and is of importance in arriving at a definite diagnosis

SYNOVIOMA

In the past the name synovioma has been applied to all forms of intracapsular tumors However the term should be applied only to a neoplasm which is derived primarily from the synovial lining of the joints bursas and tendon sheaths and is characterized by the formation within the lesion of spaces lined with synovial cells It is highly malignant Although the tumor is derived from synovial cells and may thus arise from either a joint or a bursa it may be entirely extracapsular and have no apparent association with these structures The condition is seldom recognized prior to biopsy and microscopic study The diagnosis is usually that of a calcified bursa and in most instances it is not realized that

extensive areas of bone destruction and usually show a tendency to lobulation and multiple calcific flecks Giant cell tumors of the tendons are most common in the digits Synovial cysts usually lie in the joint, are spherical, and have a smooth, sharp border

PIGMENTED VILLONODULAR SYNOVITIS

A condition which may produce manifestations closely similar to those in synovioma has been described by Jaffe and Lichtenstein under the term of pigmented villonodular synovitis It has also been designated by the names giant cell tumor of the synovia chronic hemorrhagic villous synovitis benign synovioma xanthoma and xanthogranuloma The lesion may be circumscribed or diffuse with the synovial membrane generally thickened, pigmented and covered with villous and coarse nodular outgrowths It is not thought to be neoplastic, rather appears inflammatory in nature The x ray appearances of synovial sarcoma and pigmented villonodular synovitis are identical and it is impossible to establish a definite dagnosis by roentgen study alone However the following points are suggested as being helpful (1) a lesion originating in the synovial tissues and subsequently invading bone is not pigmented villonodular synovitis and should be designated as synovial sarcoma (2) a sharply defined soft tissue mass in or near a joint and containing scattered and irregular deposits of amorphous lime will usually prove to be sarcoma, (3) in the presence of an irregular soft tissue mass extrinsic to the joint capsule with no intracapsular fluid or synovial proliferation, the lesion should be considered synovial sarcoma or other neoplasm rather than villonodular synovitis of the pigmented variety It is important to determine whether a new growth adjacent to the joint is actually within the joint By careful roentgen techniques and complete and careful study of the roentgenogram a decision can be made in many instances The clinical history and the physical examination are very important in diagnosis and the roentgenologist must not hesitate to avail himself of this data

Whereas it was previously believed that the condition was a neoplasm histologic studies indicate that the process is entirely inflammatory It is most frequently found in young adults There is usually a history of monarticular swelling persisting for months or years with no associated fever or local signs of inflammation There may be slight or moderate pain with limitation of motion and stiffness On physical examination there is an effusion into the joint which is greatly out of proportion to the patients complaints Palpable nodules may be present in the suprapatellar, infrapatellar or popliteal spaces The nodules are most common in the late stages of the disease and are more apparent after aspiration of the joint Fluid removed from the synovial space is amber or xanthochromic with the physical characteristics of a transudate The fluid reaccumulates rapidly after aspiration and the relief of pain is only temporary

The diagnosis rests mainly on clinical grounds supported by surgical confirmation The roentgen manifestations are contributory and are important to rule out involvement of the bone or other joint lesions The absence of bone changes differentiates the lesion from other diseases of the bones and joints Pneumography may show no evidence of synovial disease

disease or fifteen or more years after the onset Metastases may involve the lung, the pleura, and the lymph nodes

Roentgen Manifestations On roentgen examination, the synovioma presents as a solitary, spherical or ovoid area of homogeneous density The density is slightly greater than that of the adjacent soft tissues The margins of the soft tissue mass are smooth and sharply defined Synoviomas vary in size from 1 cm to 10 or more cm Osteoporosis is a frequent concomitant Effusion, periosteal reaction, and bone destruction are not common The synovioma is usually a homogeneous, structureless mass without dense septa or areas of increased radiance as in the lipomas The uniformity of the density is an important diagnostic feature The ease with which the lesion can be identified on the roentgenogram is influenced largely by whether the margin is demarcated by a more radiolucent zone such as the suprapatellar fatty tissue or whether it is surrounded by a mass of muscle which is essentially of the same density Soft

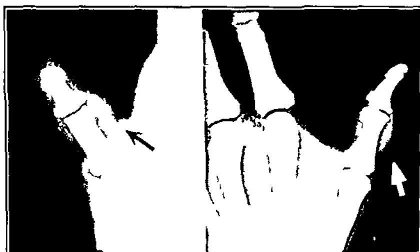


Fig 263 Synovioma A soft tissue mass is visualized in the proximal portion of the thumb (arrow) The mass is sharply defined smooth in outline and of uniform density throughout Operation revealed a synovioma

tissue techniques and multiple views are important In some instances it is not possible to outline the entire periphery of the mass Calcification in the tumor is rare and is usually amorphous irregular, and in the form of multiple, small flecks Bone destruction may occur The extent of the bone involvement is slight in comparison with the size of the tumor mass The edges of the area of destruction are irregular poorly defined and present no marginal sclerosis or thickening Serial roentgenograms may show gradual increase in the size of the mass although the tumor may remain stationary for months or years

Differential Diagnosis Less common neoplasms of the soft tissues comprise lipomas hemangiomas and glomus tumors Liposarcoma is dissimilar to synovioma and it does not occur near a joint, usually has extensive calcification and presents a heterogeneous internal pattern with multiple areas of radiolucence With fibrosarcoma there is usually extensive bone reaction areas of periosteal activity, and calcific flecks Myxosarcoma of tendon sheath origin and plexiform neuroma do not occur near joints Giant cell tumors of the tendons closely resemble synovioma They may be in intimate relationship to the joint but tend to have more

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MYOSITIS OSSIFICANS FIBROSITIS OSSIFICANS OSSIFYING FIBROSITIS

In myositis ossificans, new bone formation occurs in intimate relation to the muscles The ossification assumes shapes analogous to the muscle bundles and most commonly is adjacent to the shafts of the long bones but does not extend to the extreme end of the bone or the joint Various theories as to the cause of this condition have been advanced It is most probably consequent upon an injury with hemorrhage into the soft parts with subsequent ossification Proliferation of fragments of periosteum or bone which have been separated by trauma may also be a causative factor The site of the injury remains tender and painful much longer than usual After an interval of two to three weeks an area of calcification becomes demonstrable in the tissues The calcium deposits are arranged in lamellæ or striations parallel to the shaft and usually lie in contact with the bone at one or more points There is no destruction of the bone or perpendicular striæ as in osteogenic sarcoma With the passage of time, the calcific depositions become more extensive in size and increase in density Rest and limitation of use are indicated to diminish the extent of the calcification Early operation is usually followed by prompt and extensive recurrence Late operative removal can usually be performed safely

The term *myositis ossificans* is incorrect as the essential pathology is not a myositis and there is no actual ossification of the muscles the calcific depositions being in the connective tissue about the muscles The affected connective tissue in many instances is attached to or lies in contact with the bone The condition should be termed *fibrositis ossificans*

Myositis Ossificans Progressiva Progressive Ossifying Fibrositis

Myositis ossificans progressiva or progressive ossifying fibrositis as it should more properly be termed, is characterized by skeletal anomalies and ossification of the connective tissue in regions adjoining the bones It should be included among the lesions which are inheritable

Etiology The etiology of myositis ossificans progressiva is unknown Cases have been recorded of the occurrence of the condition in monozygotic twins The lesion has been reported in a father and son by Borton Fanning and Vaugen Gaster has described the disease in a father, grandfather and three sons of the same family Uehlinger was of the opinion that it is probably hereditary and that commonly advanced etiologic factors such as trauma, inflammation, trophoneurotic influences and blastomatous changes could be rejected The disease must be considered a true mutation of the mesenchyma It is frequently associated with deformities of the extremities which on roentgen examination closely resemble osteochondrodystrophy, a known inheritable condition

Clinical Manifestations The condition is more frequent in males The onset is usually in infancy or early childhood with a soft tissue swelling most commonly in the cervical or upper dorsal region The swelling is painful and tender and may show redness and heat In some instances the soft tissue swelling disappears spontaneously and the acute manifestations subside a firm mass remaining in the soft tissues Roentgen examination shows ossification in the mass There is gradual increase in stiff-

ness and limitation of motion of the neck, chest, back and extremities particularly the shoulders The dorsal region of the trunk is more apt to be affected than the ventral aspect and in most instances the upper half of the body more than the lower half The lower extremities are rarely involved The patient becomes thinner and weaker and chronic invalidism develops gradually with resultant disuse atrophy of the muscles During the third decade of life, there is usually spontaneous arrest Most of those afflicted die during the third or fourth decade of intercurrent infection probably due to interference with respiration In many instances

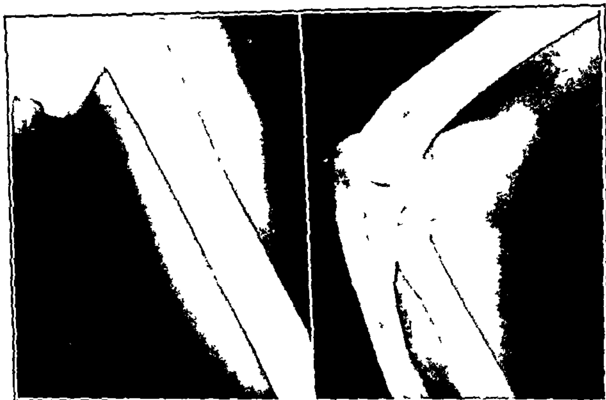


Fig 264 Fig 265

Fig 264 Myositis (Fibrositis) Ossificans The patient a male eighteen years of age had suffered a blow on the arm six weeks previously Roentgen study immediately after the injury revealed no abnormalities The calcific deposition in the soft tissues showed a gradual increase in size and density at two subsequent examinations made at intervals of three months

Fig 265 Myositis Ossificans There is an irregular area of calcific density in the soft tissues of the anterior aspect of the elbow The bones of the elbow reveal no abnormalities

there are associated congenital anomalies of the thumbs and great toes the changes being those of microdactylia with absence or shortening of some of the phalanges and fusions of the phalanges There may also be deformities of the proximal ends of the phalanges and the distal ends of the metacarpals

Roentgen Manifestations There is progressive ossification of the muscle sheaths the tendons and the intervening connective tissues Irregular band like depositions of calcific density occur in various portions of the body The common sites in which the calcifications are found comprise the cervical region particularly the sternocleidomastoid area the thoracic wall and the axillas There is extensive calcification of the ligaments of the spine In many instances there is ossification in the region

of the pelvis, the hip joints and the thighs The calcific depositions may form synostoses or false joints with the ribs the spine, the shoulder girdle or the hips and may cause bony ankylosis The density of the extra skeletal bone varies widely It may be less dense and less sharply defined than normal bone In other cases it is more dense The skeleton becomes osteoporotic due to severe limitation of motion The shafts of the bones show subperiosteal thickenings Projections similar to exostosis formations develop Fusions of the vertebrae develop in the advanced cases Fairbank describes an instance in which there was such extensive formation of bone in the quadriceps muscles as to suggest two femoral shafts and another case in which there was a continuous sheet of bone from the quadriceps to the tibia resulting in complete fixation of the knee

The microdactyly is associated with reduced length of the phalanges of the great toe In rare instances there is shortening of a metatarsal bone There is incomplete development or suppression of the proximal phalanx, this phalanx being reduced to a wedge shaped fragment In some instances, the phalanges are fused The first metatarsal may show increase in length with fusion of part of the proximal phalanx This results in the formation of a rounded end to the bone which is quite dissimilar to the normal head A similar shaping of the head may also be seen when the bone is shortened In the thumb all three bones may be short or the shortening may be limited to the phalanges In a case reported by Burrows in 1933 the metacarpal was short and the epiphysis of the proximal phalanx was wedge shaped Spurs on the back of the os calcis or anteriorly to its tuberosity have been reported There have been described exostosis formations on the phalanges of the fingers The disease shows apicocaudal progression Scoliosis develops with marked deformity of the thorax due to fixation of the ribs Bony exostoses may project from the cortices of the long bones In many instances there is defective ossification about the metatarsophalangeal joints of the great toes with marked hallux valgus similar to that which occurs in certain forms of hereditary osteochondrodystrophy

Pathology On histopathologic examination of a mass of bony structure from the soft tissues the appearance in many instances is that of normal bone The microscopic changes vary according to the stage of the disease at the time the examination is made

Diagnosis Prior to the development of bone in the soft tissue masses, diagnosis is difficult The co existence of microdactyly determines the diagnosis In calcinosis universalis the calcific deposits occur in the skin and subcutaneous tissues as well as in the muscles and in the fascia In this condition the calcific depositions are granular and fragmentary and are distinct from the shadows cast by the bony skeleton This also occurs in the case of dermatomyositis with calcification In calcinosis the depositions are frequently absorbed while in myositis ossificans progressiva the bone once formed never disappears In dermatomyositis the lesions involve the extremities and the trunk is not affected until the later stages There is fever and sweating and the spleen may be enlarged The diaphragm intercostal muscles and palate are involved It appears that dermatomyositis and myositis ossificans are closely related

SCLERODERMA

Scleroderma is a systemic disease which affects primarily the collagenous connective tissues of the skin subcutaneous tissues, and certain organs The

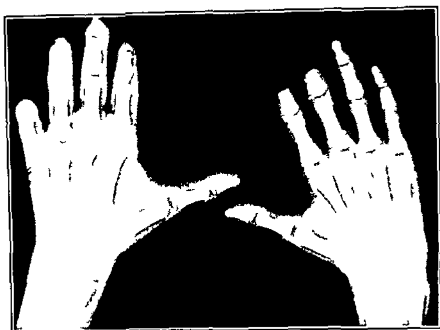


Fig 266 Scleroderma There is partial absorption of the terminal phalanges of the fingers The little finger of the right hand shows complete absence of the terminal phalanx and partial absorption of the middle phalanx Similar changes are present in the index and middle fingers on the left The soft tissues of the affected fingers are decreased in size and appear markedly irregular in outline There is osteoporosis of the bones of the hand The interphalangeal joints of the fingers show narrowing haziness and irregularity of outline

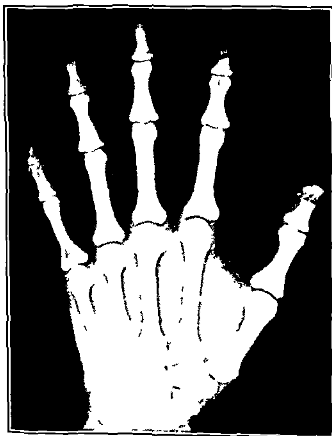


Fig 267 Raynaud s *Disease* There is partial absorption marked irregularity of outline and fragmentation of the terminal phalanges of the fingers The soft tissues of the distal portions of the fingers are denser than normal and there are multiple calcific depositions in the soft tissues of the thumb

of the pelvis, the hip joints, and the thighs The calcific depositions may form synostoses or false joints with the ribs, the spine, the shoulder girdle or the hips and may cause bony ankylosis The density of the extra skeletal bone varies widely It may be less dense and less sharply defined than normal bone In other cases, it is more dense The skeleton becomes osteoporotic due to severe limitation of motion The shafts of the bones show subperiosteal thickenings Projections similar to exostosis formations develop Fusions of the vertebrae develop in the advanced cases Fairbank describes an instance in which there was such extensive formation of bone in the quadriceps muscles as to suggest two femoral shafts and another case in which there was a continuous sheet of bone from the quadriceps to the tibia resulting in complete fixation of the knee

The microdactyly is associated with reduced length of the phalanges of the great toe In rare instances there is shortening of a metatarsal bone There is incomplete development or suppression of the proximal phalanx, this phalanx being reduced to a wedge shaped fragment In some instances the phalanges are fused The first metatarsal may show increase in length with fusion of part of the proximal phalanx This results in the formation of a rounded end to the bone which is quite dissimilar to the normal head A similar shaping of the head may also be seen when the bone is shortened In the thumb, all three bones may be short or the shortening may be limited to the phalanges In a case reported by Burrows in 1933 the metacarpal was short and the epiphysis of the proximal phalanx was wedge shaped Spurs on the back of the os calcis or anteriorly to its tuberosity have been reported There have been described exostosis formations on the phalanges of the fingers The disease shows apicocaudal progression Scoliosis develops with marked deformity of the thorax due to fixation of the ribs Bony exostoses may project from the cortices of the long bones In many instances there is defective ossification about the metatarsophalangeal joints of the great toes with marked hallux valgus similar to that which occurs in certain forms of hereditary osteochondrodystrophy

Pathology *On histopathologic examination of a mass of bony structure* from the soft tissues the appearance in many instances is that of normal bone The microscopic changes vary according to the stage of the disease at the time the examination is made

Diagnosis Prior to the development of bone in the soft tissue masses, diagnosis is difficult The co existence of microdactyly determines the diagnosis In calcinosis universalis, the calcific deposits occur in the skin and subcutaneous tissues as well as in the muscles and in the fascia In this condition the calcific depositions are granular and fragmentary and are distinct from the shadows cast by the bony skeleton This also occurs in the case of dermatomyositis with calcification In calcinosis, the depositions are frequently absorbed while in myositis ossificans progressiva the bone once formed never disappears In dermatomyositis the lesions involve the extremities and the trunk is not affected until the later stages There is fever and sweating and the spleen may be enlarged The diaphragm intercostal muscles and palate are involved It appears that dermatomyositis and myositis ossificans are closely related

SCLERODERMA

Scleroderma is a systemic disease which affects primarily the collagenous connective tissues of the skin subcutaneous tissues and certain organs The

cranial ossification The skin is extremely distensible and elastic and the joints show marked hyperextensibility Slight trauma produces large, gaping wounds which result in extensive linear or irregular scars Pseudo tumors occur over various prominences of the body, especially the elbows and knees These are due to extravasation of blood after slight injuries the hematomata undergoing partial absorption and leaving behind excessive scar tissue There is in some instances a slight increase in the excretion of 17 ketosteroids in the urine, increase in the ratio of uric acid to creatinine in the blood and eosinophilia

The condition is of special interest to the radiologist because of the fact that the patient presents peculiar subcutaneous nodules immediately beneath the skin These are apparently caused by localized avascular degeneration of fatty tissue with subsequent repair The roentgenogram reveals disseminated small, rounded, discrete, calcareous nodules which appear to involve the skin or the immediate underlying tissue They are most common on the extensor surfaces In the lower legs there is wide distribution along the lateral and medial surfaces of the lower extremity, the changes occurring in the region of the fatty tissues and where there is a special liability to trauma Calcifications usually occur as central zones of increased radiolucency surrounded by a ring like shadow of greater density In some cases the calcific deposits are stippled or homogenous They closely resemble the phleboliths found in venous angiomas While the calcifications in angiomas and the Ehlers-Danlos syndrome are quite similar roentgenographically, the conditions are easily differentiated clinically Phleboliths are apt to be situated deep in the soft tissues and present concentric strata of calcium The Ehlers Danlos nodules are superficial and involve only the extremities The larvae of parasites occur in the muscles and not in the subcutaneous tissues Roentgen study of the joints reveals increased range of motion in the joints which in some instances is very marked There may be shortening and lack of development of the proximal phalanges of the fingers although this is not a constant characteristic of the disease

SUPRASPINATUS TENDINITIS PERITENDINITIS CALCAREA OF THE SHOULDER PERIARTHRITIS CALCAREA OF THE SHOULDER

Supraspinatus tendinitis is one of the most common causes of shoulder pain The condition is known by a number of names the chief of which are subacromial or subdeltoid bursitis, peri or para arthritis of the shoulder peritendinitis calcarea, calcifying tendinitis, and the supraspinatus syndrome The symptoms comprise pain and disturbance of function related to the shoulder joint although the lesion is extrinsic to the joint At approximately the age of fifty years practically all individuals develop a certain degree of degenerative change in the supraspinatus tendon area Relatively early aging processes occur in this region most probably on a mechanical basis and associated with man s transition to the upright position Similar tendogenetic lesions occur elsewhere in the body particularly in the triceps tendon adjacent to the olecranon process the rotators distal to the head of the radius the flexors of the fingers, the tendon of the gluteus the tendon of the adductor magnus, and the tendon of Achilles

cause is unknown. The skin may be edematous, indurated, smooth, waxy, and tight, due to collagen fibre thickening and proliferation. Histologically, the stiffness and hardness of the involved tissues is explained by focal fibrinoid degeneration and diffuse sclerosis of the intradermal and subcutaneous connective tissue. The changes may be widespread involving the blood vessels, endocardial and subendocardial connective tissue, myocardium, lungs, and portions of the gastrointestinal tract, particularly the esophagus and small bowel. Weakness, weight loss, and arthralgias may be prodromal symptoms of the disease. The cutaneous involvement passes through several stages from brawny edema to a smooth, tight, waxy skin, which is not movable over the deeper tissues. Any cutaneous area may be involved; most frequently changes begin on the extremities, cheeks, and bridge of the nose, forehead and chest. There may be slow progression to involve all the skin of the face and trunk. Raynaud's phenomena and sclerodactylia are frequent while arthralgia is less frequent. Diffuse calcinosis of the skin (Thibierge-Weissenbach syndrome) is uncommon. The end result of the disease is marked subcutaneous and muscle atrophy, thinning and tightening of the skin and a progressive decrease in joint motion. Creatinuria, hyperglobulinemia and hypoalbuminemia occur. An important manifestation is widening of the periodontal space, particularly about the posterior teeth. The widening includes the roots and is distinguished from other disorders of the pericementum by its uniformity. Actual thickening of the periodontal membrane to two to four times normal has been observed. The involved teeth do not become loose.

In the early stages there is moderate periarticular demineralization of the long bones of the hand. The joint spaces in the hands and fingers show narrowing, the changes being similar to those in the rheumatoid types of arthritis. There may be marginal punched out osseous defects or extensive absorption of the phalanges. Minute, scattered calcific depositions occur in the soft tissues of the fingers. Sand-like or plaque-like calcinosis, mostly about pressure points such as the fingers, elbows and ischial tuberosities, has also been described. In the advanced stages calcification occurs in the skin, subcutaneous tissues and muscles in the form of granules and plaques. The deposits are most often found in the distal portions of the fingers and toes but may also occur elsewhere in the body. The diagnosis is best made by the presence of associated changes in the terminal phalanges of the hands and feet, the bones of these regions being decreased in size due to partial or complete absorption. There may also occur increased density of the phalanges and synostosis of the distal two phalanges.

EHLERS-DANLOS SYNDROME

The Ehlers-Danlos syndrome is a congenital dystrophic anomaly or constitutional dyscrasia of the mesenchyma with certain hereditary or familial tendencies. It is characterized by specific manifestations in the skin, joints and subcutaneous tissues with hyperelasticity and fragility of the skin and blood vessels, hypermobility of the joints, pseudo tumors over the bony prominences, weakness, osteoporosis, delayed epiphyseal development, and moveable nodules beneath the skin. Other congenital anomalies are frequently present, particularly synostosis of the proximal ends of the radius and ulna, club feet, supernumerary teeth, and delayed

cranial ossification. The skin is extremely distensible and elastic and the joints show marked hyperextensibility. Slight trauma produces large gaping wounds which result in extensive linear or irregular scars. Pseudo tumors occur over various prominences of the body, especially the elbows and knees. These are due to extravasation of blood after slight injuries, the hematomata undergoing partial absorption and leaving behind excessive scar tissue. There is in some instances a slight increase in the excretion of 17-ketosteroids in the urine, increase in the ratio of uric acid to creatinine in the blood and eosinophilia.

The condition is of special interest to the radiologist because of the fact that the patient presents peculiar subcutaneous nodules immediately beneath the skin. These are apparently caused by localized avascular degeneration of fatty tissue with subsequent repair. The roentgenogram reveals disseminated small rounded discrete calcareous nodules which appear to involve the skin or the immediate underlying tissue. They are most common on the extensor surfaces. In the lower legs there is wide distribution along the lateral and medial surfaces of the lower extremity, the changes occurring in the region of the fatty tissues and where there is a special liability to trauma. Calcifications usually occur as central zones of increased radiolucency surrounded by a ring-like shadow of greater density. In some cases the calcific deposits are stippled or homogenous. They closely resemble the phleboliths found in venous angiomas. While the calcifications in angiomas and the Ehlers-Danlos syndrome are quite similar roentgenographically, the conditions are easily differentiated clinically. Phleboliths are apt to be situated deep in the soft tissues and present concentric strata of calcium. The Ehlers-Danlos nodules are superficial and involve only the extremities. The larvæ of parasites occur in the muscles and not in the subcutaneous tissues. Roentgen study of the joints reveals increased range of motion in the joints, which in some instances is very marked. There may be shortening and lack of development of the proximal phalanges of the fingers, although this is not a constant characteristic of the disease.

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Supraspinatus tendinitis is one of the most common causes of shoulder pain. The condition is known by a number of names, the chief of which are subacromial or subdeltoid bursitis, peri- or para-arthritis of the shoulder, peritendinitis calcarea, calcifying tendinitis and the supraspinatus syndrome. The symptoms comprise pain and disturbance of function related to the shoulder joint, although the lesion is extrinsic to the joint. At approximately the age of fifty years practically all individuals develop a certain degree of degenerative change in the supraspinatus tendon area. Relatively early aging processes occur in this region, most probably on a mechanical basis and associated with man's transition to the upright position. Similar tendogenetic lesions occur elsewhere in the body, particularly in the triceps tendon adjacent to the olecranon process, the rotators distal to the head of the radius, the flexors of the fingers, the tendon of the gluteus, the tendon of the adductor magnus and the tendon of Achilles.

Calcific depositions in the tendons about the shoulder are common and in routine chest surveys it has been demonstrated that they occur in approximately 3 to 5 per cent of the population In many instances, they produce no pain or limitation of motion In others the encapsulated mineral deposit serves as a focus of a sterile inflammatory reaction The adjacent tissues become swollen and hyperemic the overlying subacromial bursa loses its lubricating function and the calcific deposition increases in size, in some instances becoming sufficiently large to interfere with motion by impinging against the acromion process and the coraco acromial ligament There is pain on active and passive motion of the shoulder Without treatment the process persists for several weeks leaving residuals of atrophic, contracted muscles, a shrunken joint capsule, shortened ligaments and adhesions in the region of the subacromial bursa

Anatomy A bursa is a sac lined with synovial membrane Normally, it is collapsed and contains a small amount of synovial fluid The bursas occur at points of motion between adjacent parts The subacromial or subdeltoid bursas lie between the deltoid muscle and the acromial arch superiorly and the superior portion of the shoulder joint capsule and the greater tuberosity of the humerus inferiorly They do not communicate with the shoulder joint In some instances the subacromial bursa is not a single unit, there being a delicate partition partially or completely separating the subdeltoid and subacromial divisions It may also have a subcoracoid extension The function of the bursa is to facilitate movement for the greater tuberosity of the humerus under the acromion in abduction of the shoulder It also is important in rotation of the humerus The bursa fails to function properly in the presence of inflammation, fibrosis or calcification In acute inflammation hydrops develops and is manifested by acute swelling and pain The fibrosis may be so marked that the sac is completely obliterated The subacromial bursa which is slightly smaller than the palm in each individual, fits like a cap over the lateral aspect of the shoulder between the outer and inner muscle planes Small calcifications frequently are found within the short rotator tendons and periosteal proliferations occur at the site of the insertions of these muscles into the humerus Small bone fragments may become separated from these insertions

Pathologic and Clinical Findings The lesion originates as a degenerative process in the tendon fibers usually in the supraspinatus portion of the cuff Avascular necrosis occurs in the tendon fibers and is aggravated by inefficient mechanics attrition occupational strain, and the processes of aging In some instances there are degenerative changes and varying degrees of inflammation without calcific deposition while in others there are extensive calcifications in the tendons or bursa without associated symptomatology

The calcium salts are in the form of carbonates and phosphates the composition usually being about 20 per cent calcium carbonate and 80 per cent calcium phosphate Calcific depositions are most common in the region of the shoulder but have also been found in the elbow wrist, hand, hip knee and foot and about the trochanters of the femur and the coracoid process Studies of the calcium phosphorus and alkaline phosphatase content of the blood reveal no abnormalities The larger areas are more apt to cause symptoms and the size of the deposit is in general proportionate to the clinical manifestations The term calcified bursitis is widely used in the medical literature with reference to the painful shoul-

der in which calcific depositions are demonstrable in the roentgenogram It is important to stress that the disease arises primarily in degenerated tendon rather than in the bursa, the alterations in the bursa being secondary It is incorrect to speak of calcification of the subacromial or subdeltoid bursa or of calcified subacromial or subdeltoid bursitis Since the bursal changes associated with actual calcific depositions in the supraspinatous tendons are secondary rather than primary, calcific tendonitis peritendinitis calcarea or periarthritis calcarea are more representative and should replace the older terminology which is actually incorrect and misleading



Fig 268 Fig 269

Fig 268 Calcified Tendinitis Right Shoulder There is an extensive area of calcific density in the soft tissues overlying the head and tuberosity of the humerus calcification of the tendon and bursa

Fig 269 Peritendinitis Calcarea of the Shoulder There is a large irregular area of calcific density overlying the tuberosity and the adjacent portion of the head of the humerus

Roentgen Technique Calcifications in the soft tissues are frequently found on roentgen study of the shoulder girdle Fractures with separation of small spicules of bone are common There may also occur injuries to the tendons such as rupture of the supraspinatus tendon and dislocation of the long head of the biceps brachii The roentgenologist is frequently asked to localize as accurately as possible these and other abnormalities in the shoulder The usual anteroposterior views of the shoulder with the hand in supination and pronation frequently do not suffice to establish the diagnosis Stereoscopic study often is not helpful By taking a series of 5 roentgenograms it is possible to bring the site of each muscle insertion into profile in at least two planes and localize defects involving the periosteum and cortex and calcifications or bone fragments in the soft tissues The first view is taken with the shoulder elevated on a 23 degree angle board with the arm externally rotated and the palm turned upward It serves to open the subacromial space between the head of the humerus

and the acromion of the scapula, brings into profile the tendon insertion of the infraspinatus muscle, and demonstrates calcifications which otherwise are overshadowed by the head of the humerus For the second position, the angle board is removed, the arm and forearm being maintained in external rotation The summit of the superior facet for the insertion of the greater portion of the fibers of the tendon of the supraspinatus muscle on the greater tuberosity is brought into profile In the third position, the patient is supine with the arm and forearm rotated internally and the palm turned down on the table top This brings into view the facet for the insertion of the teres minor tendon on the posterior aspect of the greater tuberosity This position may be modified by placing the patient prone with the arm in internal rotation and the forearm flexed behind the back The fourth view is taken with the patient seated The arm and forearm are abducted 90 degrees from the side The internal and external epicondyles at the lower end of the humerus are palpated and the plane of axis rotated to right angles with the horizontal The central ray is directed medially and cephalad towards the patient s ear with the head erect This brings the long axis of the acromial and coracoid processes along with the glenoid neck of the scapula into view The acromioclavicular and shoulder joints are visualized at right angles to the previous projections The surface of the lesser tuberosity of the humerus is rotated into profile anteriorly and demonstrates the insertion for the subscapularis tendon The fifth view is termed the bicipital The patient is supine the arm is externally rotated to the fullest extent and the elbow is held as close to the body as possible The tube is brought below the level of the table top, directed toward the head and angled approximately 10 to 15 degrees The central ray is directed into the length of the bicipital groove This gives an excellent view of the groove and throws the tuberosities into relief

Roentgen Manifestations The roentgen diagnosis is established by the demonstration of an area of calcific density in the soft tissues overlying *the head and tuberosity of the humerus The size varies from a barely* visible fleck to a very large area measuring several centimeters The density may be granular and mottled or uniform The position of the calcified mass is variable One of the commonest sites is directly adjacent to the lateral border of the tuberosity of the humerus Other common locations are in the region of the neck and upper portion of the shaft of the humerus overlying the head of the humerus and below the acromion process A less frequent location is in the soft tissues below the inferior margin of the glenoid The borders of the area of calcific density *may be smooth and sharply defined or irregular in outline There is in* many instances roughening of the lateral margins of the tuberosity of the humerus Osteoarthritic changes with spurring about the margins of the acromioclavicular joint are of frequent occurrence The bones of the shoulder may show osteoporosis This is usually not marked in degree and is most pronounced in the upper third of the humerus The soft tissues about the shoulder are increased in thickness and density and there is loss of the definition of the subcutaneous and muscular layers in the acute phases of the disease The chronic stage may be associated with atrophy of the musculature It must be stressed that the calcific depositions may be demonstrable only in certain projections and not in others hence a complete study utilizing all of the positions described above may be required to establish the diagnosis The presence of a fracture is ex

cluded by the absence of a defect in the bones of the shoulder. Other conditions which must be considered in differential diagnosis include primary or metastatic neoplasm of bone, osteomyelitis, tuberculosis and neurotrophic arthropathy. None of these lesions causes the manifestations which characteristically occur in peritendinitis calcarea and the diagnosis in most instances is established without difficulty.

Treatment. The treatment varies with the severity of the symptoms. Common methods comprise diathermy, manipulation under general anesthesia, needling with or without irrigation and operative removal of the deposit. Roentgen treatment has proven highly effective. It usually results in relief of pain and re-establishes freedom of motion. In most instances 4 to 6 roentgen treatments are administered at intervals of two to four days to the affected shoulder joint. The therapy is less effective in the chronic type. The earlier it is instituted, the more satisfactory is the response. In many instances the first and second treatments do not give relief and in some cases the pain is intensified after the first treatments. Reabsorption of the calcium deposition occurs in a large percentage of the cases.

DEPOSITION OF CALCIUM NEAR THE ELBOW

Deposits of calcium in the common extensor tendon of the forearm are unusual. Roentgen study reveals a dense, irregular calcific shadow adjacent to the epicondyle. In patients treated by early operation the area of density is found to be due to the deposition of a viscous fluid consisting largely of calcium phosphate in the region of the origin of the common extensor tendon. In differential diagnosis, it is necessary to consider loose bodies and osteoarthritis, conditions which are distinctive for baseball and tennis players and have commonly been termed the "tennis elbow." Rarely there may be lipping of the ulna at its articulation with the internal condyle. The most distinctive lesion is the development of single or multiple extra-articular deposits of bone in the ligamentous and tendinous attachments beneath the ulnar nerve. Removal of the deposits usually affords relief.

CALCIFIC DEPOSITIONS IN THE WRIST AND HAND

Calcific depositions are rare in the wrist and palm. In a series of cases reported by Seidenstein there were 259 instances with involvement of the shoulder and only 2 cases of calcific deposits at the wrist. The clinical picture is usually characterized by severe local pain. The roentgen examination establishes the diagnosis by demonstration of the shadow of the calcific deposition in the soft parts. The calcification may be homogeneous, measuring only 3 to 4 mm. in diameter and rounded in shape. In other cases it is extremely large and dense, measuring as much as 1 × 2 cm. The very small lesions are easily overlooked if the condition is not suspected. The opaque areas may be long, linear and fragmented. The exact location of the depositions is speculative as surgical exploration is seldom utilized. It appears that the larger depositions are much too extensive to be confined within the substance of any tendon and doubtless lie within the peritendinous tissues such as the tendon sheaths, the bursas or the ligaments.

It is believed that the calcification is asymptomatic and lies in a relatively avascular and insensitive tendinous and ligamentous structure with subsequent rupture of the deposit into the sensitive synovial or bursal tissues. The cases have shown an equal distribution as regards sex. The ages in the reported cases vary from thirty to sixty nine years, the largest group being in the forties. It has been noted that many of the patients show a calcific deposit in the shoulder simultaneously or shortly afterward.



Fig 270. Peritendinitis Calcarea of the Wrist. There is an irregular area of mottled calcification in the soft tissues of the anteromedial aspect of the wrist in the region adjacent to the pisiform. The patient had pain and tenderness in the wrist.

PELLEGRINI STIEDA'S DISEASE (POST-TRAUMATIC CALCIFICATION OF THE COLLATERAL TIBIAL LIGAMENT OF THE KNEE)

Pellegrini Stieda's disease is a post traumatic calcium deposit in the region of the collateral tibial ligament of the knee and manifests itself in the roentgen examination as an area of calcification lying in the soft tissues medial to the inner condyle of the femur. The shadow of the calcified mass does not become visible on the roentgenogram until two weeks or more after the injury. It may therefore be necessary to make repeated roentgen studies to establish the diagnosis. Pellegrini published the first case of this disease in 1905 and in 1908 Stieda made an apparently independent record of a similar case. Pellegrini Stieda's disease is post traumatic in origin. The injury may be severe or slight and may be a direct blow to the knee or an indirect trauma such as tripping or falling. The disease may result from repeated slight injuries. In no case has Pellegrini Stieda's disease occurred spontaneously as the result of infection, or in conjunction with a neoplasm of the knee.

Pathology Pellegrini Stieda's disease is a deposition of calcium in the soft tissues overlying the medial condyle of the femur The calcification is in the collateral tibial ligament This ligament is a flat band applied to the medial side of the knee joint, it arises from the inner epicondyle of the femur distal to the adductor tubercle and passes downward attaching to the medial condyle and medial portion of the shaft of the tibia below the level of the tuberosity Microscopically the calcified mass of

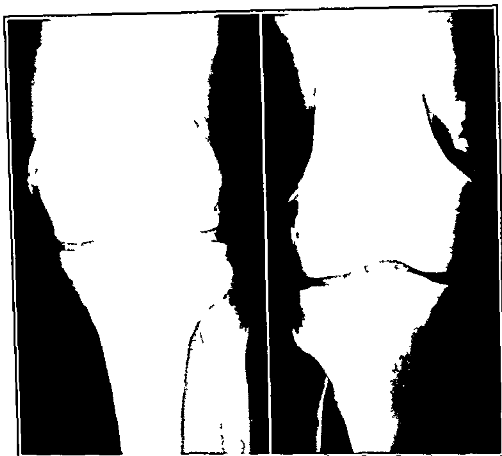


Fig 271 Fig 272

Fig 271 Pellegrini s Disease There is a lunate shaped area of calcific density in the soft tissues overlying the medial condyle of the femur calcification of the collateral tibia ligament

Fig 272 Pellegrini Stieda s Disease There is a large irregular area of calcific density in the soft tissues adjacent to the medial aspect of the lower end of the femur calcification in the collateral tibial ligament The patient suffered a trauma to the knee eight months previously

this disease is definitely bony in character and infiltrates the tendons and ligaments overlying the medial aspect of the knee The bony mass is composed of three types of bone growth (1) primitive bone on the basis of connective tissue (2) callus like bone and (3) enchondral variety of cartilage ossification

Signs and Symptoms In the period directly following the injury, the symptoms are those associated with trauma to the knee namely pain swelling tenderness and limitation of motion The pain and disability persist longer than would be expected with the trauma received and it is this failure of the symptoms to subside in the usual time which leads to further roentgen studies after an interval of several weeks to months

The calcified mass may be palpable, it is hard, usually slightly movable, and is not attached to the skin overlying it

Roentgen Manifestations The roentgen examination reveals an area of calcification in the soft tissues medial to the inner condyle of the femur The shadow is usually crescentic but may be elongated rounded or triangular in shape The dimensions of the abnormal shadow are variable The length varies from 0 5 cm to 8 0 cm and the thickness from 2 mm to 1 5 cm As the disease progresses, the shadow usually enlarges in all diameters The density of the calcified area changes with the age and size of the lesion Recently formed masses cast a slight shadow, sometimes so faint as to be barely visible After the lapse of a few weeks, it becomes increasingly opaque and when it is of months' or years standing the calcified area becomes as dense as the femur The shadow is usually of uniform density throughout its entire extent The borders of the calcified mass are in most instances sharply defined and smooth in outline, at times, especially in cases of long duration, the borders are irregular, serrated and poorly defined The typical shadow is entirely free in the soft tissues overlying the medial condyle of the femur and there is an area of radiance between it and the femur This is of great importance in ruling out the possibility of osteoma periostitis or fracture of the femoral condyle as the cause of the abnormal density There may be a fracture periosteal tear, or similar condition in conjunction with Pellegrini Stieda s disease In rare instances there appears to be a zone of calcification connecting the shadow in the soft tissues with the femoral condyle, this may be caused by the angle at which the roentgenograms are taken The soft tissues about the area of calcification are thickened and denser than normal

The calcified area does not make its appearance until about three weeks after the trauma to the knee although it has been reported as early as fifteen days If the patient is examined soon after the injury there is swelling and thickening of the soft tissues about the inner aspect of the knee but no demonstrable abnormality Many patients are thereupon considered as presenting merely a strain or sprain and the correct diagnosis is therefore overlooked If the possibility of Pellegrini Stieda s disease is borne in mind the patient is advised to return in two or three weeks at which time a faint thin area of density is visualized in the soft tissues Subsequent examinations reveal the typical findings described above Anteroposterior roentgenograms demonstrate the shadow best In the oblique position the calcified area may appear to be attached to the femur because of overlapping In the lateral projection the lesion usually is obscured by the femur

CALCIFICATIONS OF THE BURSAS OF THE KNEE

There are multiple bursas about the knee the principal ones being the prepatellar the infrapatellar and the superficial pretibial over the insertion of the patellar ligament There are also variable bursas below and above the collateral ligaments and numerous popliteal bursas Calcification of the bursas about the knee is often not suspected until revealed by roentgen examination It is an important cause of symptoms and must always be considered in the differential diagnosis in patients with pain, tenderness and limitation of motion involving the knee

POST-TRAUMATIC PARA-ARTICULAR CALCIFICATIONS AND OSSIFICATIONS ABOUT THE ANKLE

The diagnosis of para-articular calcification about the ankle in many instances can be established only by serial roentgen studies At the time of the injury the roentgen examination reveals no bone involvement, there being only soft tissue thickening and infiltration about the joint The triangular dark shadow which is present above the calcaneus posteriorly, the supracalcaneal triangle, may be encroached upon or obliterated as a result of hemorrhage or exudate which may be intracapsular, extracapsular or both Re-examination after three to four weeks usually shows the deposition of calcium The calcifications parallel the surface of the posterior malleolus of the tibia, the superior and anterior aspect of the talus or may lie along the external margin of the malleolus There is usually a definite clear space between the calcification and the bone The calcific particles may lie between the fibula and the tibia The shape and size of the calcification are variable Most commonly it is crescent shaped However many are linear Calcification of a hematoma is usually of rather diffuse appearance and the deposits are irregular, amorphous, and confined to the mass of the hematoma The calcific depositions may ossify and show a trabecular structure throughout the mass In the late stages there may be a fusion between the ossified density and the adjacent bone The changes are analagous to those in post-traumatic calcification of the collateral tibial ligament of the knee

CALCIFICATION OF THE MENISCI OF THE KNEE

There are two types of calcification of the menisci, primary and secondary In the primary type calcium is laid down in parallel layers throughout the entire meniscus Usually more than one meniscus is affected There is roughening and enlargement of the meniscus The primary type is the result of a nutritional deficiency occurs with advancing age and is a degenerative change It occurs in association with calcification of the supraspinatus tendons and the trochanteric bursas of the femurs The secondary type results from trauma and is found more frequently in younger individuals The calcification may affect both menisci of the knee and in some instances is bilateral The condition is manifested by mild pain in the knees Locking of the knee with severe pain is not infrequent Bloody effusion may develop Partial dislocation of the meniscus may occur Localized swelling and limitation of motion are usually present Inflammation, redness and heat are generally absent Slight twisting of the knee initiates pain Calcifications of the semilunar cartilages of the knee are analagous to calcification in the connective tissue and fibrocartilage of other joints of the body

SYNOVIAL OSTEOCHONDROMATOSIS

Synovial osteochondromatosis also termed synovial chondromatosis is characterized by the development of osteocartilaginous bodies in the synovial membrane and their extrusion into the joints and adjacent soft tissues as loose bodies They remain pedunculated

and attached to the synovia The joints most commonly involved are the hip, knee, and shoulder Trauma and infection aggravate the lesion but do not appear to be the cause The bodies are demonstrable only when calcified or containing bone, the cartilaginous type



Fig 273 Calcification of the Semilunar Cartilages There is extensive calcification of the semilunar cartilages manifested by linear and plaque like areas of calcific density overlying the articular surfaces of the tibia



Fig 274 Synovial Chondromatosis The multiple areas of mottled calcification in the infrapatellar region are characteristic of synovial chondromatosis

being demonstrable only by means of injection studies. A common site of involvement is the hip. A significant roentgen manifestation is the presence of a defect at the junction of the head and neck of the femur. The neck of the femur presents a conical shape. There may be areas of erosion in the acetabulum and the head of the femur. Loose bodies are frequently present and are associated with areas of resorption in the bone. The sites of the localized areas of erosion in the hip joint are due to the distribution of the cartilaginous bodies at the junction of the synovia and the cartilage. Involvement of the shoulder produces similar changes in the humerus and scapula. In the knee, there may be erosions of the femur and tibia. The loose bodies may erode through the capsule and lie outside of the joint. When there are only a very small number of opaque foreign bodies, arthrography is of aid in diagnosis. It is particularly valuable in the differentiation of loose body formations due to other causes such as osteochondritis dissecans. In the case of the knee, the multiple small opaque bodies may lie posteriorly or anteriorly in the region below the patella.



FIG. 275. Synovial Chondromatosis. *A*, Anteroposterior projection. The calcific shadows overlie the neck and upper end of the shaft of the femur and are diffusely distributed in the soft tissues. *B*, Lateral projection. The calcific deposits surround the upper end of the femur.

ARTHROGRAPHY

Spontaneous Occurrence of Gas in Joints Vacuum Pneumarthrography

Gas is not infrequently present in the joint spaces and may be of clinical significance Traction applied to an extremity may produce a decrease of pressure in the joint with release of gas from the adjacent tissues into the joint space In the hip joint, the spontaneous vacuum phenomenon may make it possible to demonstrate irregularity of the cartilage which otherwise could not be visualized Gas may be present in the soft tissues in the "bends" The gas is a mixture of oxygen, carbon dioxide and nitrogen similar to that in the circulating blood It may be possible by aid of the contrast afforded by the gas in the knee joint to demonstrate a lesion of the meniscus, loose bodies, and deformities of a cartilage associated with arthritis Gas may occur in the sacroiliac joints and may cause confusion in diagnosis The so called vacuum sign is occasionally visualized in various joints, particularly in the knee hip and shoulder upon the application of strong traction In the sacroiliac joint or intervertebral disc it may be a manifestation of a disease process In the case of the knee, spontaneous vacuum may persist for several weeks It is caused by shrinkage and constriction of the meniscus It may be bilateral, is usually associated with pain, and is unrelated to trauma The occurrence of gas in joints generally is not of pathologic significance Clinically, it is manifested as 'cracking of the knuckles ' It is common in the shoulder joints of children whose hands are being held over the head for immobilization in making chest x rays After about ten minutes the gas is replaced by fluid Passive torsion of the spine may result in the accumulation of gas in the apophyseal joints in the lumbar spine Absence of the vacuum space in joints under traction or other circumstances in which it normally should occur may be of diagnostic significance in that it is indicative of fluid in the joint

Internal Derangements of the Knee Joint Vacuum Technique

The structures demonstrable on the roentgenogram of the knee are the skin the subcutaneous layer with its large veins the rectus femoris and the vasti the suprapatellar space the patellar ligament, the infrapatellar fat pad and its extensions the tendons of the gastrocnemius and hamstring the adductor muscle and its tendon the cartilage lining of the joint the tibial collateral ligament the extracapsular bursas when distended and the menisci While soft tissue studies are helpful they are not essential The examination must include anteroposterior lateral and oblique views Valuable data can be obtained by a longitudinal view of the patella and intercondylar projections A special light source which can be regulated to vary not only the intensity but also the size of the field of illumination and the shape of the opening is invaluable for optimum visualization of the soft tissue structures

The internal meniscus may be demonstrated by use of the so called vacuum technique utilizing traction or abduction of the leg This is accomplished by placing the patient in the lateral recumbent position with a sandbag between the table and the outer aspect of the knee to be studied A horizontal roentgen ray beam is directed anteroposteriorly through the knee while an assistant forces the leg into abduction against

the table top The exposure is made about two or three seconds after the initiation of the traction or abduction If made earlier or later, full vacuum is not obtained due to incomplete dispersion or filling of the joint space with synovial fluid A lateral view of the meniscus is obtained by the same method Slight irregularity of the margins or small defects in the menisci are not of significance and are not uncommon in healthy knee joints The internal meniscus can be demonstrated in about 80 per cent of the cases while the external meniscus can be visualized in not more than 20 per cent If the meniscus in the injured joint cannot be shown while the meniscus in the healthy knee is demonstrated the findings indicate the presence of increased amounts of synovial fluid in the injured knee, even though there is no swelling or other clinical manifestation of fluid If the meniscus in both knees cannot be demonstrated the findings are of no value The demonstration of a slight increase in the synovial fluid in an acutely injured knee as evidenced by failure of demonstration of the meniscus emphasizes the importance of a careful search for abnormal changes in the soft structures It is essential to exclude the presence of fracture or loosening of the attachment of the meniscus, particularly in the presence of swelling of the tibial collateral ligament The structures in the region of the tibial collateral ligament are visualized in the anteroposterior and lateral views A rupture or sprain of the collateral tibial ligament is manifested by localized swelling of the ligament Demonstration of the meniscus in a swollen knee indicates the absence of an excess of synovial fluid and indicates that the swelling may be due to subcutaneous edema, enlargement of the bursas periosteal damage hemorrhage, a torn ligament a cyst, or a hypertrophied fat pad The attachments of the menisci vary widely

Loose bodies may be single or multiple pedunculated or free, laminated or non laminated They may originate from unorganized fibrin necrotic portions of the synovial membrane organized connective tissue cartilage or bone Hemorrhage into the joint may be the source of fibrin which subsequently develops into a loose body Necrotic synovia in association with an arthritic process may form loose bodies This is particularly common in tuberculosis Loose bodies may originate from hypertrophied synovial fringes due to trauma, synovitis, osteoarthritis, and hypertrophied fat pads Many chondromas are of synovial origin Tabes produces large fragments of bone Non tuberculous types of infectious arthritis usually produce small bony sequestra

The tibial collateral ligament is a broad, flat membranous band situated nearer the back of the joint Swelling of the ligament due to contusion or edema is slight while in the presence of hemorrhage it is apt to be marked There is usually an associated synovitis and the increased synovial fluid prevents demonstration of the meniscus Swelling or thickening of the injured ligament is best demonstrated by comparison with the opposite knee In chronic sprain or rupture thickening of the ligament and fibrosis in the overlying subcutaneous tissues may be present In rare instances ossifying periostitis occurs

The bursas about the knee are normally not visualized When distended with fluid as the result of bursitis they may be demonstrated Swelling of the suprapatellar bursa is most easily shown as the bursa is easily distensible and may contain large amounts of fluid The prepatellar bursa is visualized when it contains even small amounts of fluid In Osgood Schlatter s disease there may be swelling of the pre tibial

bursa Cysts of the posterior bursa or synovial membrane are best visualized on soft tissue roentgenograms The cysts may arise from any bursa but are most common in the posterolateral aspect of the knee On extension and flexion of the knee, the cyst changes in size and shape, an important aid in diagnosis The cysts are degenerative in nature and are comparable to ganglia They arise in the synovial and extrasynovial fatty and areolar tissues and fibrocartilage and are most common in the external meniscus Tumors of the soft tissues comprise xanthomas angiomas, and fibrolipomas, and may be visualized by careful soft tissue studies Varicosities of the popliteal and long saphenous veins can be outlined by roentgen methods Contusions with extracapsular edema or hemorrhage may be distinguished from intracapsular swelling by visualization of the meniscus

Pneumarthrography of the Knee

In pneumarthrography it is essential that the contrast medium have a high degree of safety with a minimum of irritation and be rapidly absorbed An opaque medium has certain advantages and iodine compounds as used in excretion urography either alone or in combination with oxygen or air have been utilized The use of opaque media has not proven satisfactory They may obscure bone details or loose bodies and cannot be removed completely in all instances with the resultant possibilities of irritation, foreign body reaction and adhesions They may cause a flare-up of an old infection There have been several cases of myositis ossificans reported after the use of iodized oil All opaque media previously used in the study of the joints have caused deleterious reactions Air carbon dioxide and oxygen have been utilized by many observers the last being considered the medium of choice The oxygen should be dust free and sterile It has the advantage of being non irritating and is absorbed more rapidly than air Carbon dioxide has proven equally satisfactory The possibility of embolism must be borne in mind If direct entry of the gas into a blood vessel is avoided oxygen may be used with impunity Many thousands of injections have been done without sequelae and no deaths have been reported There is danger of emboli in intra articular fracture with injury to the veins and arteries and of dissemination of infection in the presence of suppurative arthritis The method is contraindicated in hemophiliacs and in patients with ankylosis

The Anatomy of the Knee The knee is a shallow double hinged joint situated between the two largest and strongest bones in the body As with the spine the knee has assumed a new role in the upright position The weight bearing portions of the knee are protected by two semilunar cartilages The menisci deepen the sockets of the tibial condyle or the plateau The medial meniscus is C shaped Its inferior periphery is attached to the tibia by the coronary ligament The remainder of the face of the meniscus is fused with the capsule to the inner portion of the medial collateral ligament forming a simple and complete attachment The lateral meniscus is nearly circular Its attachment is similar except that there is no peripheral attachment to the joint capsule at the point at which it is crossed by the popliteal tendon and muscle In cross section the meniscus is triangular in shape and is attached at its base with a free inner margin Normally the capacity of the synovial cavity is 80 to 120

cc It is incompletely divided into an anterior portion two posterior joint sacs and a large suprapatellar pouch One or more popliteal bursas may communicate with the knee joint

Technique Strict aseptic technique is essential Procaine is infiltrated into the skin A needle is inserted under the patella through the capsule and synovia into the joint This may be accomplished from the lateral or medial side The patella is displaced manually toward the side of injection in order to localize the sulcus between the patella and the femur Excessive fluid is withdrawn The gas is injected slowly into the joint through a syringe until the synovia is distended and the patient feels a sense of fullness and discomfort in the knee In the adult male 80 to 120 cc is usually necessary The volume of gas utilized must be noted to determine the capacity of the joint The amount is diminished in certain types of arthritis adhesions and chronic synovitis After the injection has been completed and the needle removed pressure is applied firmly over the puncture for several seconds This prevents subcutaneous emphysema which produces confusing extraneous shadows and may necessitate repetition of the study at a later date The knee is flexed gently to distribute residual fluid around the menisci and into the suprapatellar pouch and to spread the medium through the joint cavity Air may be retained in the joint for several days Carbon dioxide and oxygen are absorbed rapidly

Roentgen Technique It is important that the joint space be localized accurately the joint spaces spread and roentgenograms taken at various angles to permit of visualization of all parts of the joint Traction is exerted on the thigh and ankle by mechanical methods or by hand Sachs and his co workers recommend that seven roentgenograms be made, a lateral and six in the postero anterior projection Three views are made with the medial aspect of the joint spread (1) internal rotation with the foot inverted (2) a neutral position with the foot pointed down and (3) with the foot in external rotation or the foot everted Similarly, three views are taken with the lateral portion of the joint spread A lateral projection is then made without traction Cardboard holders with high speed film Bucky diaphragm extension cone distance of 36 inches 72–80 kv and 200 milliampere seconds are used

The Normal Pneumarthrogram The structures which may be visualized are the menisci the synovial sacs, the infrapatellar fat pad the cruciate and articular ligaments, and the articular cartilage The femoral tibial and patellar cartilages measure about 2 or 3 mm in thickness and extend along the margins of the bones The lower border of the femoral articular cartilage is outlined as a curvilinear density adjacent to the condyles On the medial aspect this linear shadow overlies the outer two-thirds of the density produced by the medial meniscus while laterally it overlies the inner third of the shadow of the lateral meniscus The semilunar cartilages are two wedge shaped structures which possess a slight degree of mobility and change in size and position as the joint is moved The lateral meniscus is closely related to the intercondyloid eminence and is separated from the collateral fibular ligaments by the popliteus tendon It is sharply outlined along its superior and inferior surfaces The popliteus bursa is continuous with the knee joint and gas in the bursa may simulate a tear of the lateral meniscus In the lateral projection, the anterior aspect of the meniscus is continuous with the infrapatellar fat pad anteriorly and with the posterior portion of the articular capsule posteriorly The

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bodies both opaque and nonopaque can be established by pneumoarthrography. It is possible to demonstrate articular and bony changes in acute and chronic arthritis, osteochondritis dissecans, chondromalacia and osteochondrosis of the patella. It is invaluable as a preoperative aid to the surgeon. In careful hands and with proper techniques the method is entirely safe and is not followed by any complications. It is a simple and practical means of diagnosis of internal derangements of the knee joint and is particularly valuable in cases which cannot be diagnosed by other means.

Diodrast Arthrography of the Knee

Diodrast may be used in place of air or oxygen as a contrast medium for arthrography. There is sharp contrast which may bring certain pathologic changes within the realm of accurate preoperative diagnosis. There are no subjective reactions. Sections of synovia removed surgically reveal no microscopic evidence of reaction to the contrast medium. Ten cc. of 35 per cent solution is injected into the knee joint and the limb is manipulated gently to spread the medium over the joint surfaces. Anteroposterior, postero-anterior, lateral and oblique stereoscopic roentgenograms are taken. Films exposed one half hour after the injection normally reveal only a small residual of the opaque medium. The method is of importance in cases of old or recent trauma and may reveal tears of the menisci, ruptures of the anterior cruciate ligaments and Baker's cysts.

posterior cruciate ligament is outlined in the posteroanterior roentgenogram as a broad, oblique band of density within the intercondyloid fossa. The anterior cruciate is superimposed upon the lateral border of the posterior cruciate shadow due to the fact that it arises from the anterior intercondyloid fossa between the anterior horns of both menisci and extends posteriorly toward the lateral condyle.

The collateral ligament is adherent to the medial meniscus. The fibular collateral ligament is separated from the lateral meniscus by the tendon of the popliteus. The ligaments are outlined by the gas adjacent to their medial margins and are smooth in outline. The infrapatellar fat pad is triangular in shape. It is visualized only in the lateral projection and occupies the anterior tibial condylar space, extending to the anterior wall of the capsule and the patella. It lies outside the joint and is extrasynovial.

The bursas about the knee joint are divided into an anterior and posterior group. The anterior bursas are three in number. The suprapatellar bursa lies between the femur, the quadriceps extensor tendon and the patella, while the infrapatellar bursa is situated between the tibia and the patellar ligament at the inferior margin of the fat pad. Both these bursas are clearly outlined on the pneumogram. The prepatellar bursas lie between the skin and the patellar ligament. They do not communicate with the joint and usually are not visualized. The posterior bursas are divided into three main groups. In the lateral group are the gastrocnemius bursa and the semimembranous bursa. Other bursas occur at the insertion of the collateral tibial and fibular ligaments. The articular capsule is attached posteriorly to the intercondyloid fossa of the femur and to the condyle of the tibia. The quadriceps tendon, the patella and the patellar ligament lie anteriorly and the oblique popliteal ligament lies posteriorly. The synovial membrane lines the capsule and communicates with the suprapatellar and infrapatellar bursas and the sheaths of the popliteus and gastrocnemius bursas.

Roentgen Manifestations of Lesions of the Knee Joint. Tears or fractures of the semilunar cartilages may be single or multiple. There may be gross infiltration of the gas beneath the medial meniscus. Elevation of the medial meniscus is abnormal. The lateral meniscus may be elevated by the gas under normal conditions. The apron-like appearance of the overlying femoral cartilage reflected over the meniscus may closely simulate a fracture line. The popliteal tendon synovial sheath communicates with the joint cavity and may be reflected over the outer surface of the lateral cartilage under normal conditions and must not be confused with a fracture. A diagnosis of dislocation of the meniscus is established by elevation of the cartilage and extensive infiltration of gas between the cartilage and the cartilaginous surfaces of the tibia. Post-operative changes in the knee joint following removal of the meniscus are manifested by absence of the shadow of the normal cartilage. Injuries to the cruciate ligaments such as tears, calcification and hypertrophy of the ligaments are demonstrable. The anterior cruciate ligament is more apt to be torn than the posterior. Hypertrophies and calcifications of the infrapatellar fat pads may be demonstrable. Loose bodies within the knee joint may be the result of synovial osteochondromatosis, osteochondritis dissecans, hypertrophic arthritis and chip fractures. Chronic synovitis is characterized by thickening of the synovial membrane. Baker's cyst can be outlined in the pneumarthrogram. The exact localization of foreign

A Physical,
 1 Thermal or radiation effects
 2 Chemical,
 3 Vascular (ischemic),
B Traumatic
C Idiopathic (angioneurotic groups)

Thermal Osteosis In gangrene of an extremity subsequent to burning or freezing the affected skeletal parts become necrotic The necrotic areas are demarcated and become separated from the intact portions of the bone by osteoclasis and proliferation of granulation tissue Bone has less resistance to low temperatures than soft tissue since after the freezing of an extremity, extensive necrosis of bone may develop while the muscles, vessels nerves and skin remain intact Also the epiphyseal cartilage becomes necrotic while the articular cartilage remains unaffected The differences in resistances of the epiphyseal and articular cartilage are due to differences in the sources of nutrition Epiphyseal cartilage receives its blood supply from the adjacent bone while the articular cartilage is nourished by diffusion from the synovial fluid Injury to or necrosis of the bone marrow lessens or interrupts the flow of nutritive elements to the epiphyseal cartilage whereas the nutrition of the articular cartilage is not affected

Radiation Osteosis Necrosis of bone after irradiation with radium or roentgen rays is well known Because of its marked susceptibility to irradiation bone can be destroyed by dosages which leave the soft tissues covering the bone still intact The increased susceptibility of bone to irradiation is important in the administration of roentgen or radium therapy (see pp 458-470)

Chemical Osteosis Chemical osteosis today is rare Formerly it occurred in dentistry after the application of arsenic and in factories which utilized phosphorus in the manufacture of matches Arsenic applications have been discontinued by the dental profession and the use of yellow phosphorus in factories is unlawful The aseptic necrosis usually involved parts of the interdental septum, particularly in those cases in which the application of arsenic to a dental cavity was not followed by hermetic sealing

Ischemic Osteosis The occlusion of blood vessels by an embolus or arteriosclerosis results in varying degrees of necrosis of the bones which are deprived of their blood supply The extent and delimitation of the necrosis are dependent on the presence of collateral circulation In most instances conditions are not favorable for the establishment of a collateral circulation because the arteries which supply the bone are relatively small

Traumatic Osteosis Trauma leads to necrosis of bone when it destroys or markedly restricts the blood supply of the bone In consequence, necrosis of bone is a common sequel to fractures as the injury results in the rupture of many blood vessels Necrosis of isolated fragments of bone is frequent The development of necrosis in parts of the main fragments of the fractured bone is dependent on the relation of the fracture to the blood vessels which supply the bone In adults injury to the nutrient artery or one of its branches may result in necrosis of large areas of the bone marrow with secondary necrosis of adjacent spongy or compact bone Fractures in the area of the metaphysis are more favorably situated

ASEPTIC NECROSING LESIONS OF BONE

Aseptic necrosis, also termed subchondral necrosis or osteochondritis, is a term used to designate a group of conditions characterized by localized necrosis of the epiphysis, the metaphysis, or a short bone The sites at which the lesion may occur and the names of those who first described the condition are the tubercle of the tibia (Osgood Schlatter), the tarsal navicular (Kohler), the epiphysis of the head of the femur (Calve, Legg, Perthes), the head of the second metatarsal (Freiberg), vertebral epiphyses (Scheuermann), the sternal end of the clavicle (Friedrich), the head of the humerus (Lewin), the internal condyle of the humerus (Legg, Lohr) the olecranon process and the greater and lesser trochanters of the femur (Monde Felix), the head of the metacarpal bones (Mauclaire), the iliac crests (Buchman), the pubis (Van Neck) the patella (Larsen), the upper end of the tibia (Ritter) the lower end of the tibia and the fibula (Stern), the talus (Diaz) the epiphysis of the first metatarsal (Wagner) the carpal navicular (Preisser) the carpal lunate (Kienbock), the distal end of the femur (Konig) the calcaneus apophysis (Sever Haglund)

The etiology is usually uncertain and is not always the same in the various forms of the disease The most generally accepted theory is that the lesion is caused by interference with the blood supply to the part or bone affected with resultant anemia and necrosis In many cases there is a history of trauma or repeated mild traumas Other possible causes which have been suggested comprise sepsis tuberculosis and disturbances of the sympathetic innervation Males are much more commonly affected probably due to greater activity and more frequent liability to trauma The term 'osteitis has been applied collectively to all of the pathologic lesions in this group However it should not be used to designate a non-inflammatory degenerative bone change as the suffix conforming to this lesion is ' osis which signifies 'a condition of' or "a state caused by ' *Common usage has differentiated the suffixes " itis" and " osis "* nephritis indicating inflammation of the kidney while nephrosis indicates a non inflammatory or degenerative disease of this organ

The reactions of the bone tissue in aseptic necrosis are in some respects similar to those which occur after an inflammatory process involving bone During the resorption of segments of necrotic bone there occur simultaneously proliferation of young connective tissue and the formation of numerous new blood vessels When the processes of regeneration, resorption and formation of bone continue undisturbed, the osteitis may heal completely Mechanical injuries or secondary infection interfere with the process of healing and the lesion may result in permanent deformity of the affected bone Secondary infection may be due to invasion of bacteria from the surface The infection may also be hematogenous in origin, the bacteria involving the necrotic area because of its lowered resistance With the development of secondary infection the aseptic necrosis becomes a septic process and may accurately be termed an osteitis The term osteosis should be used to designate the group of conditions known as the aseptic necroses of bone and the designation "osteitis' should be applied only when secondary infection has set in

The following is suggested as a classification of the various forms of osteosis

and is frequently subjected to spontaneous or pathologic fractures. A fracture always traverses the necrotic area and does not occur between the living and dead bone. At this stage, the idiopathic osteosis becomes evident by roentgen methods. The continued friction between the fragments results in a grinding of the necrotic bone and the zone of fracture is soon filled with bone dust. An area of density is created in one part of the necrotic bone and becomes separated from the adjacent bone by a barrier which offers resistance to resorption and regeneration of the peripheral walled off bone. While regeneration of the proximal zone continues, the barrier itself is gradually removed and is replaced by a layer of young connective tissue. In consequence, a separation takes place between the regenerated and the necrotic part of the damaged bone. This stage is clearly demonstrable in the roentgenogram. The affected area is sharply demarcated and is covered to a greater or lesser extent by the damaged articular cartilage. The necrotic fragment may in some instances unite with the regenerated fragment. The involved bone may show a permanent division, particularly in the carpal, tarsal and sesamoid bones. In most cases in which the smaller parts of the articular condyle or head of the bone become necrotic, as in osteochondritis dissecans, the nonunion of the necrotic fragment and the rest of the bone may result in the formation of free bodies within the joint. The necrotic segments may, after an initial period in which they show increased radiance due to osteoclasis, enter a stage of greater radiopacity from impregnation of the necrotic tissue with calcium salt.

OSTEOCHONDRITIS OF THE CARPAL LUNATE (KIENBOCK'S DISEASE)

OSTEOCHONDRITIS OF THE CARPAL NAVICULAR (PREISSER'S DISEASE)

Kienbock in 1910 published the first description of the clinical symptomatology and roentgen appearance of the disease which is now known by his name. Osteochondritis of the carpal navicular (Preisser's disease) is similar in its etiology and manifestations, hence these lesions are discussed together. The condition occurs chiefly in the right or working hand of young adult manual workers. The lesion progresses through three stages, the onset often being subsequent to a slight injury and resembling a sprain with slight pain which persists for days or a few weeks. The second stage is characterized by a period in which the patient is free from symptoms and persists for a few months. In some instances an additional injury causes the abnormality to be discovered at this time. In the third phase there is usually reactivation of the disease, the symptoms continuing for years and terminating in marked osteoarthritis of the wrist joint. The etiology is uncertain. The fact that the lunate bone is most commonly affected suggests that the explanation may be based upon the peculiar anatomy of the bone. The greater part of the surface of the lunate is covered by articular cartilage. There are two small areas on the anterior and posterior surfaces which have a periosteal covering and through which blood vessels enter the bone. The principal blood supply

with relation to the nutrition because the blood supply to the metaphysis and epiphysis is from a nutrient artery on one side, while the arteries which enter through the articular capsule and ligaments supply the other aspect of these structures.

In many instances a trauma may not be sufficient to produce a fracture and yet, especially if repeated many times, results in disturbance of the blood supply to the bone at a vulnerable point. The effect varies with the type of bone and the portion of bone affected. When the involved blood supply is to the cancellous bone, slow disintegration results. After the lapse of several months the bone disintegrates and collapses. Subsequently revascularization takes place and repair is brought about by fibrous tissue replacement of the dead bone, organization, and new bone formation. In some instances the stimulus to repair is not sufficient to permit of complete repair and areas of necrosis persist. These appear as small radiant cystic areas on the roentgenogram, as in the diseases of Freiberg, Preisser and Kienbock. If the affected blood supply is from a small end artery in the subchondral portion of a joint, a small area of infarction develops and deprives a fragment of the articular cortex of nutrition. This occurs in osteochondritis dissecans. When the area of involvement affects the blood supply to the metaphyseal region of a growing epiphysis, necrotic changes develop. An epiphysis which is subjected to severe stress may undergo detachment and slipping. This is particularly apt to occur in the epiphysis of the proximal end of the femur.

PATHOLOGY. On histopathologic study there are four stages: (1) primary subchondral necrosis. There are no roentgen manifestations at this time; (2) thickening and rarefaction of the bone structure. The normal bony outline is preserved. Occasionally there is sequestrum formation, indicating that the bone is undergoing necrosis. The unaffected bone appears osteoporotic while the necrotic bone is of normal density; (3) in the third stage bone deformity develops. The structure is not uniform, there being multiple zones of increased and decreased density, often with *pathological fractures. There are evidences of resorption of necrotic bone* and simultaneous new bone formation as evidenced by layers of granulation tissue with osteoclasts and osteoblasts about the foci of necrotic bone; (4) there is reformation of normal bone accompanied by marked deformities. The resultant deformities together with the changes in the cartilage result in the development of arthritis deformans in the late stages.

Idiopathic osteosis is a disease of childhood and adolescence, particularly if the epiphyses are involved. In other parts of the body, notably the carpal and the metatarsal bones, it may occur in later years, most commonly in the twenties or thirties. It appears that the sequence of events is characteristic for all these conditions and the development of the lesions is similar for the entire group. The early stage is a necrosis involving the bone tissue and marrow of an epiphysis or part of the epiphysis, or of a bone or part of a bone. On gross and roentgen examination no changes are demonstrable at this time. The second stage is that of repair. During resorption of the necrotic bone, young connective tissue invades the involved area and new bone is substituted for the necrotic bone. The regenerative process may, if undisturbed, result in complete healing. The necrotic bone loses its normal resistance to mechanical stress

pain limited motion, and swelling aggravated by use Because of the deformity of the bone the adjacent joints are subjected to abnormal stresses and chronic arthritic changes develop with increasing disability

Prognosis The outlook should always be guarded, complete restoration of function seldom taking place The duration is prolonged and the disability and pain persist despite operation in most cases

Differential Diagnosis The differential diagnosis must include a large number of lesions which may produce closely similar changes In fracture, there is a history of sudden, severe trauma with x ray evidence of a fracture Partite navicular is congenital in origin, is often bilateral and produces no symptoms as a rule *In osteomyelitis there is a history of* sudden onset with fever, chills marked local swelling redness and pain Chronic arthritis involves several bones and is usually bilateral Primary or metastatic neoplasm is rare in the carpal bones

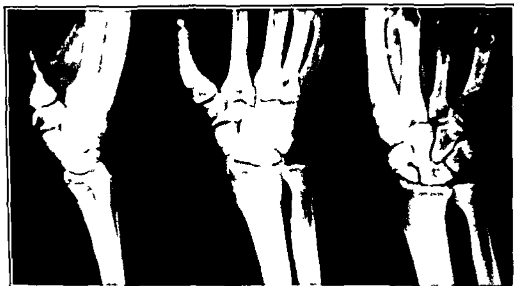


FIG 277 Osteochondritis of the Navicular Preisser s Disease There is an old ununited fracture of the navicular with slight separation of the fragments The navicular shows cystic changes increased density mottling and irregularity of outline indicative of osteochondritis The lunate shows cystic lesions

COXA PLANA

Coxa plana is also known as Legg Perthes disease Calve s disease osteochondritis deformans juvenilis, and aseptic necrosis of the upper femoral epiphysis It is more common in boys than girls and is unilateral in 90 per cent of the cases The usual age of onset is three to twelve years The lesion is fundamentally a vascular disturbance involving the femoral head with subsequent degenerative changes The soft tissues about the joint become swollen edematous and hyperemic, particularly the synovial membrane and there develops perivascular lymphocytic and plasma cell infiltration There is softening and decalcification of the diaphyseal side of the disc followed by absorption and reformation of bone The capital epiphysis is deformed and there is luxation of the hip with deformity of the acetabular surface The first stage of the disease lasts several weeks and is characterized by soft tissue changes and synovitis This is followed by the degenerative or active phase which persists for about two years

is through the anterior carpal ligament, the posterior blood supply being less efficient Variations in the relative length of the lower ends of the radius and ulna are an important anatomical feature Under normal conditions, the lower ends of these bones are equal in length In some instances the ulna is shorter than the radius, while in others the reverse is true This difference in length occurs in approximately 40 per cent of normal persons Variation in the lengths of the bones may facilitate trauma by impingement of the lunate on the adjacent portion of the radius Most observers consider trauma the primary cause However, the lesion may develop as the result of embolic occlusion of the vessels It is believed that the effect of trauma is to diminish or obliterate the blood supply to the bone with resultant avascular necrosis In the differential diagnosis of the sprained wrist, Preisser's and Kienbock s diseases must always be considered A single negative roentgenogram should not be accepted as final, and it is essential to examine the patient on more than one occasion at intervals of several weeks

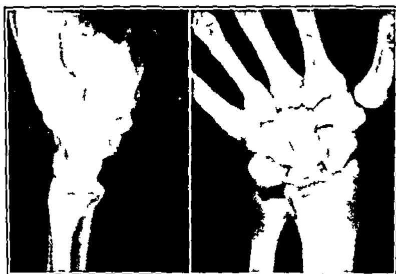


FIG 276 Osteochondritis of the Lunate Kienbock s Disease The lunate is increased in density narrowed irregular in outline and mottled

The clinical and roentgen manifestations may be divided into four stages (1) In the early or incipient stage there is pain with negative radiographic findings The duration of this phase is one to fourteen days (2) In certain instances there is absence of symptoms and the patient is of the opinion that recovery is taking place However, roentgen study at this time reveals increased density of the affected bone There may be slight narrowing of the bone The general contour and outline remain normal The duration of this stage varies from a few weeks to several months (3) After a variable interval the patient again experiences discomfort, limitation of motion and pain with swelling of the wrist X ray study shows definite and characteristic alterations with irregular calcification some areas being almost devoid of calcium while others are so dense as to suggest sequestrum The bone outline is deformed, indicative of a compression type of fracture with wedging Osteoporosis of the adjacent bones accentuates the density of the affected bone At times, a cystic type of degeneration is present The duration of this stage is one to three years (4) In the final stage there are evidences of chronic arthritis with

tion reappears The head remains mushroom in shape and subluxated The shape of the acetabulum adapts itself to the head of the femur Osteoarthritis often develops in middle life This change is usually less severe than in cases of slipping of the upper femoral epiphysis Degenerative changes similar to those in coxa plana often develop in the course of treatment of congenital dislocation of the hip slipping of the upper femoral epiphysis or fracture of the neck of the femur and are due to disturbances of the circulation of the head Coxa plana pursues a self-limited course and after a period of years usually undergoes healing There is a residual deformity in practically every instance the head and neck remaining broad flattened and shallow and the acetabulum becoming similarly deformed Early diagnosis and prompt institution of therapy are essential to prevent ankylosis shortening and loss of function Untreated cases may have marked deformity with shortening of the limb and permanent disability Arthritic changes are common in later life particularly in cases which are improperly treated

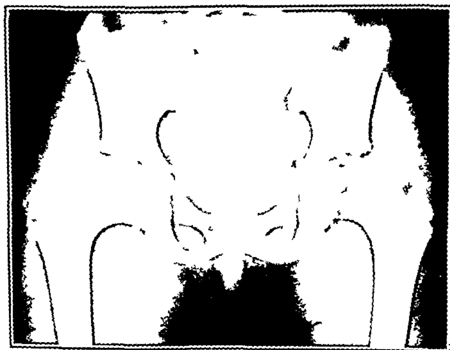


Fig. 279 Legg Perthes Disease Bilateral

OSTEOCHONDRITIS OF THE TIBIAL TUBERCLE (OSGOOD SCHLATTER S DISEASE)

The proximal extremity of the tibia is comprised of two condyles which become confluent anteriorly and form a somewhat flattened surface of triangular outline The apex of this confluence is termed the tubercle of the tibia The tubercle is divided into two parts The upper portion is rounded and smooth while the lower part is rough and receives the insertion of the ligamentum patellae The proximal tibial epiphysis ossifies at about the time of birth During early childhood the tibial tubercle develops from a tongue of cartilage which grows from the proximal tibial epiphysis and extends over the anterior aspect of the tibial metaphysis An ossification center develops in this cartilaginous projection This

Healing usually requires three to four years and the final result is the residual stage with deformity The commonest symptoms are a slight limp and pain referred to the groin thigh, and knee In the residual stage, the disability is due to faulty mechanics

It appears probable that coxa plana is not a distinct disease entity The condition may occur as a symptomatic or an idiopathic manifestation Gaucher s disease and sickle cell anemia must be included as probable causes, the simultaneous appearance of these diseases and Legg Perthe s syndrome having been noted with sufficient frequency to exclude merely a coincidental relationship In a case reported by Draznin and Singer, a female child of five years showed the characteristic manifestations of Gaucher's disease with enlargement of the liver and spleen symptomatic thrombocytopenic purpura, and anemia due to bone marrow infiltration by Gaucher s cells Roentgen examination revealed the characteristic manifestations of Legg s disease There were also multiple radiolucent



FIG 278 Legg Perthe s Disease The head of the right femur is small irregular in outline and fragmented The right acetabulum is widened and shallow and the right femoral neck is short and broad There is osteoporosis of the right side of the pelvis and the right femur

defects throughout the femoral shaft the Erlenmeyer flask appearance of the lower femur lytic lesions in the mandible and changes in the lumbar spine sacro iliac and lumbosacral joints

Roentgen Manifestations The earliest roentgen evidence of the disease is a globular swelling of the capsule The joint space is widened superiorly because of flattening of the ossified portion of the epiphysis There is no change in the shape of the articular cartilage at this stage The epiphyseal line appears wide and irregularly decalcified at its junction with the neck *Irregular* dense areas appear *in the capital epiphysis* usually *in the central* portion The dense areas becomes irregularly decalcified During the stage of decalcification the head becomes wider flatter and thinner and the neck thicker and shorter the so called mushroom appearance The soft tissue swelling subsides gradually There is marked atrophy of the affected femur and the bones of the pelvis The epiphyseal line recalcifies and the proximal surface of the neck becomes convex and extends into the head Recalcification takes place slowly and eventually normal trabecula

tion reappears. The head remains mushroom in shape and subluxated. The shape of the acetabulum adapts itself to the head of the femur. Osteoarthritis often develops in middle life. This change is usually less severe than in cases of slipping of the upper femoral epiphysis. Degenerative changes similar to those in coxa plana often develop in the course of treatment of congenital dislocation of the hip, slipping of the upper femoral epiphysis, or fracture of the neck of the femur and are due to disturbances of the circulation of the head. Coxa plana pursues a self-limited course and, after a period of years, usually undergoes healing. There is a residual deformity in practically every instance, the head and neck remaining broad, flattened and shallow and the acetabulum becoming similarly deformed. Early diagnosis and prompt institution of therapy are essential to prevent ankylosis, shortening and loss of function. Untreated cases may have marked deformity with shortening of the limb and permanent disability. Arthritic changes are common in later life, particularly in cases which are improperly treated.

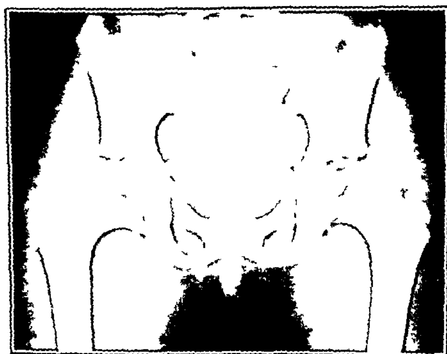


Fig. 279. Legg-Perthes's Disease. Bilateral.

OSTEOCHONDRITIS OF THE TIBIAL TUBERCLE (OSGOOD-SCHLATTER'S DISEASE)

The proximal extremity of the tibia is comprised of two condyles which become confluent anteriorly and form a somewhat flattened surface of triangular outline. The apex of this confluence is termed the tubercle of the tibia. The tubercle is divided into two parts. The upper portion is rounded and smooth while the lower part is rough and receives the insertion of the ligamentum patellae. The proximal tibial epiphysis ossifies at about the time of birth. During early childhood the tibial tubercle develops from a tongue of cartilage which grows from the proximal tibial epiphysis and extends over the anterior aspect of the tibial metaphysis. An ossification center develops in this cartilaginous projection. This

Healing usually requires three to four years and the final result is the residual stage with deformity The commonest symptoms are a slight limp and pain referred to the groin, thigh, and knee In the residual stage the disability is due to faulty mechanics

It appears probable that coxa plana is not a distinct disease entity The condition may occur as a symptomatic or an idiopathic manifestation Gaucher s disease and sickle cell anemia must be included as probable causes the simultaneous appearance of these diseases and Legg Perthe s syndrome having been noted with sufficient frequency to exclude merely a *coincidental relationship In a case reported by Draznin and Singer,* a female child of five years showed the characteristic manifestations of Gaucher s disease with enlargement of the liver and spleen symptomatic thrombocytopenic purpura, and anemia due to bone marrow infiltration by Gaucher s cells Roentgen examination revealed the characteristic manifestations of Legg s disease There were also multiple radiolucent



FIG 278 Legg Perthe s Disease The head of the right femur is small irregular in outline and fragmented The right acetabulum is widened and shallow and the right femoral neck is short and broad There is osteoporosis of the right side of the pelvis and the right femur

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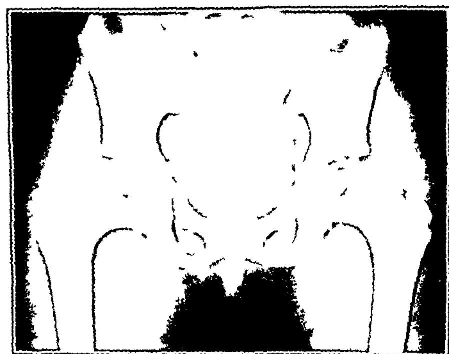


Fig. 219 Legg Perthe s Disease Bilateral

OSTEOCHONDRITIS OF THE TIBIAL TUBERCLE (OSGOOD SCHLATTER'S DISEASE)

The proximal extremity of the tibia is comprised of two condyles which become confluent anteriorly and form a somewhat flattened surface of triangular outline The apex of this confluence is termed the tubercle of the tibia The tubercle is divided into two parts The upper portion is rounded and smooth while the lower part is rough and receives the insertion of the ligamentum patellae The proximal tibial epiphysis ossifies at about the time of birth During early childhood the tibial tubercle develops from a tongue of cartilage which grows from the proximal tibial epiphysis and extends over the anterior aspect of the tibial metaphysis An ossification center develops in this cartilaginous projection This

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In osteochondritis of the tibial tubercle or Osgood Schlatter's disease there is aseptic necrosis of the tubercle. This lesion occurs in children between the ages of ten and seventeen years prior to fusion of the epiphysis. Separation and lack of fusion of the tibial tubercle can occur in adults and should not be confused with Osgood Schlatter's disease. As a result of violent exercise, there is tension of the ligament and the epiphysis with partial separation of the tibial tubercle. This results in interference with the blood supply of the epiphysis, the supply being from the patellar tendon until bony union has taken place. There is necrosis of the tibial tubercle with osteolysis and subsequent bone proliferation. The earliest roentgen manifestations comprise enlargement or thickening of the patellar ligament. Later, there develop secondary changes in the form of irregular spicules of bone. The spicules may radiate from the anterior surface of the tubercle and extend into the patellar tendon. Islands of bone may develop within the tendon and are due to ossification in the tendon or fragments which have been avulsed from the tubercle. The tubercle itself shows alteration in texture, irregularity of outline and fragmentation. The fragments are small or large and may be displaced from the shaft. In order to establish a diagnosis of osteochondritis, there must be thickening of the patellar ligament at its insertion, irregular spicules of bone extending from the anterior surface into the ligament, islands of bone in the ligament itself, and fragmentation of the tubercle with displacement of the fragments from the shaft. The disease occurs in three forms. In some cases, there is a simple epiphyseal separation. Fragmentation and necrosis may follow. A second form of the disease is an aseptic necrosis of the tibial tubercle. The third variety consists of fragmentation and necrosis of the epiphysis with, in most cases, epiphyseal separation. This is the usual form of the disease. Differential diagnosis must include pyogenic infections, syphilis and tuberculosis. These processes involve the medullary and cortical bone primarily and the epiphysis secondarily.

OSTEOCHONDRITIS OF THE PATELLA (LARSON JOHANSSON DISEASE)

In 1921 Larson and Johansson independently described a rare form of osteochondritis affecting an accessory center of ossification at the lower pole of the patella. It is an osteochondritis of the Osgood Schlatter-Kohler type. Accessory centers of ossification occur as anomalies of the patella in various sites. The commonest is at the upper lateral margin, less common are one or more lateral centers, and the rarest is an accessory center at the lower end of the patella. Osteochondritis of the inferior aspect of the patella is infrequent because of the rarity of the accessory center. The clinical manifestations may be absent or very slight. The disease occurs in children between the ages of seven and fourteen. It is more common in boys and is frequently bilateral. The lesion may occur in association with other forms of osteochondritis. The symptoms are frequently precipated by repeated mild sprains. The patient usually has a slight limp and a localized area of tenderness at the lower pole of the patella. In rare instances, the accessory center may be palpable. The disease is characterized by the development of an acute epiphysitis with interference of the blood supply and subsequent avascular necrosis. Healing is usually associated with fusion of the accessory center to the body of the patella. On roentgen examination, there is alteration of the normal

center is usually single, although in some instances it is multiple The ossification center appears usually at the age of eleven to twelve years Fusion of the tibial tubercle apophysis with the proximal tibial epiphysis occurs at about eighteen years of age On the roentgenogram, it is visualized as a beak like projection which extends downward anteriorly to the diaphysis and comprises the so called tongue shaped process of the proximal tibial epiphysis If the ossification is incomplete a separate osseous nucleus makes its appearance at the distal extremity of the lingula This is termed the anterior epiphysis and subsequently unites with the tongue shaped process The fusion of the lingula with the shaft of the tibia usually occurs at the age of eighteen years but may be delayed to the age of nineteen years

There are many variations in the development and fusion of the tibial tubercle The apophysis for the tibial tubercle may develop on one side



Fig 280 O good Schlatter s Disease There is separation and fragmentation of tibial tubercle apophysis The patient injured the knee while jumping The lesion in the upper end of the diaphysis of the tibia represents a non-osteogenic fibroma

in a different direction than on the opposite or to a variable extent on the two sides The ossification centers may develop earlier on one side In some instances an additional osseous nucleus may appear distal to the lingula Rarely there is disturbance of fusion of the tibial tubercle with the proximal tibial epiphysis or with the adjacent tibial diaphysis The patellar ligament may have a variable length of insertion and a different anatomical relationship to the tibial tubercle The patellar ligament is a continuation of the central portion of the quadriceps tendon some of the fibers being prolonged over the front of the patella to form the ligament It is a strong flat band attached above to the lower border of the patella and inserted into the lower part of the tibial tubercle and the upper part of the crest of the tibia The ligament overlies the apophysis The apophysis is in contact with the apophyseal plate and the apophyseal plate with the tibial metaphysis Roentgenographically the unfused tibial tubercle is visualized as a loose fragment of bone in the patellar ligament It constitutes a point of anatomical weakness as it results in the attachment of the patellar ligament being mechanically inadequate

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FREIBERG'S INFRACTION (OSTEOCHONDRITIS DEFORMANS METATARSO JUVENILIS)

In the so called Freiberg's infraction there is involvement of the distal end and articular surface of the second and/or third metatarsals It usually occurs in girls and young women under eighteen and there is in most instances a history of an injury with pain and disability, aggravated by use Trauma and the pressure of tight shoes causing interference

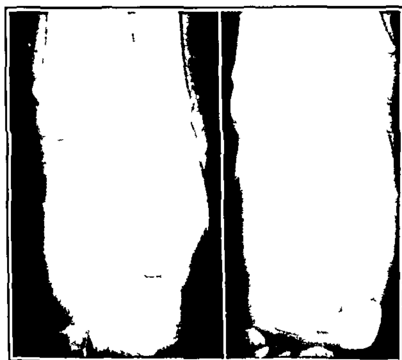


Fig 282 Kohler's Disease The right tarsal navicular is small dense and fragmented The bones of the right foot are moderately porotic

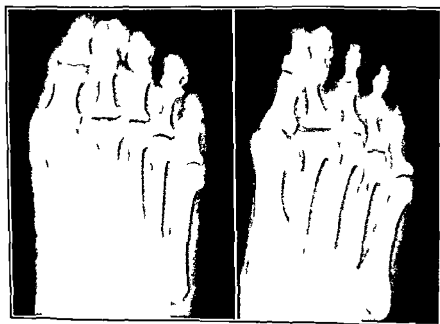


Fig 283 Osteochondritis Freiberg's Infraction The distal end of the second metatarsal is flattened widened and irregular in outline

contour and trabeculation of the inferior extremity of the patella This region may become blurred and irregular and show islands of condensation in an area of rarefied bone In differential diagnosis, it is necessary to exclude fracture, tuberculosis and other acute diseases of the knee Therapy usually consists of support In severe cases immobilization in plaster from four to six months usually results in a cure The fragments may remain permanently ununited

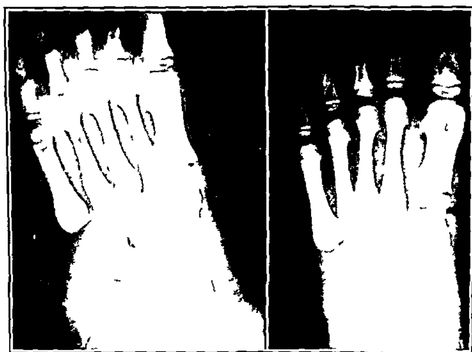


FIG 281 Kohler s Disease Osteochondritis of the Tarsal Navicular The tarsal navicular is small fragmented and mottled The patient had pain and a slight limp There was no history of trauma

OSTEOCHONDRITIS OF THE TARSAL NAVICULAR (KOHLER S DISEASE)

Kohler s disease is characterized by marked narrowing irregularity, increased density and mottling involving the tarsal navicular The condition is usually unilateral although it may be bilateral It most commonly occurs at about four to eight years While there is in some instances a history of trauma, in many others the lesion is associated with pain and disability which have developed spontaneously Swelling and limitation of motion are common In some cases there are no symptoms the condition being discovered during routine roentgen studies It may occur in association with other necrosing bone lesions indicating that it is part of a generalized bone defect or anomaly At times only the tuberosity of the navicular is involved in which case there is a separate center of ossification for the tubercle with the development of an anomalous accessory bone to which the posterior tibial tendon is attached The navicular is the keystone of the long arch of the foot Support of the arch is essential during the period of growth The disease usually disappears spontaneously whether treated or untreated and in later life the navicular usually appears entirely normal Prominence of the ridges or irregularities of outline of the adult navicular may be due to residuals from Kohler's disease in childhood

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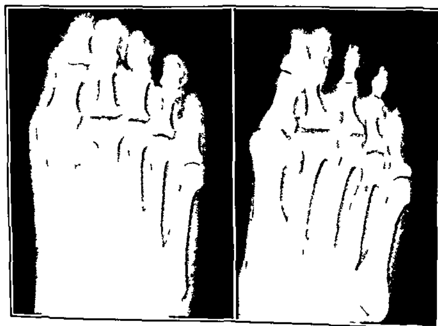


Fig 283 Osteochondritis Freiberg's Infraction The distal end of the second metatarsal is flattened widened and irregular in outline

with the blood supply are the apparent causative factors Roentgenographically, there is flattening and irregularity of the distal end of the involved metatarsal bone(s), a crescentic shape may be present in some instances The end of the bone is widened increased in density and may show small rounded, punched out areas of increased radiance The proximal end of the adjacent phalanx is usually broadened and flattened and *the metatarso phalangeal joint is widened Loose bones may occur in* the involved region Subchondral necrosis occurs with complete obliteration of the articular surface The joint is preserved

OSTEOCHONDRITIS OF THE CALCANEUS

Aseptic necrosis of the calcaneus is manifested by flattening, narrowing and mottling of the apophysis of the calcaneus A traumatism may call attention to the condition or initiate studies which reveal the lesion The trauma is not considered the causative factor On roentgen examination, the apophysis of the calcaneus reveals mottled areas of increased density with irregularity of outline, flattening and narrowing The adjacent portion of the calcaneus is roughened There is usually atrophy of the bones of the foot and ankle The process is self-limited and spontaneous recovery is the rule

JUVENILE VERTEBRAL OSTEOCHONDRITIS (SCHEUERMANN'S DISEASE)

Scheuermann s disease is also known as adolescent kyphosis or osteochondritis vertebralis juvenilis The lesion affects the thoracic spine and usually involves several vertebræ It is characterized by pain and kyphosis in the dorsolumbar region There may be tenderness over the affected area Muscle spasm is slight or absent and there is usually no wasting of the muscles Movements of the spine are limited particularly flexion The symptoms subside rapidly with bed rest and immobilization of the spine in a plaster jacket or brace

Roentgen Manifestations In patients in whom the disease occurs before the secondary centers of ossification in the epiphyseal rings have appeared the only demonstrable abnormality is wedging of the affected vertebral bodies After ossification of the rings the associated features of osteochondritis develop with irregularity of the upper and lower epiphyseal rings Schmorl s nodes and sclerosis of the adjacent vertebral surfaces The intervertebral discs between the affected vertebræ may be narrowed and this causes confusion with tuberculosis However there is no bone destruction rarefaction or abscess formation in Scheuermann s disease

Pathogenesis Adolescent kyphosis is primarily a lesion of the intervertebral discs and is associated with herniation of disc material into the vertebral bodies the so called Schmorl s nodules The herniations cause increased pressure between the adjacent vertebræ the effect being more marked along the anterior aspects of the vertebræ Roentgen changes are frequently demonstrable in the absence of symptoms The onset of the disease may be associated with vigorous exercise or heavy work The condition is not due to infection Scheuermann noted that it was similar

to Legg s disease or osteochondritis of the hip and Kohler disease of the navicular He considered it an osteochondritis or aseptic necrosis due to epiphysitis of the ring of the epiphysis which is undergoing ossification at this age At about the fourteenth year of life there appears at the superior and inferior edges of each vertebral body a small triangular shadow which represents the first sign of the epiphyseal ring The ring formations extend posteriorly in the form of fine lines and widen at the posterior edges of the vertebral bodies The epiphyseal rings remain separated from the bodies of the vertebrae until about the twenty second to the twenty fifth year of life fusion indicating that the longitudinal growth of the vertebral bodies has been completed



FIG 284 Scheuermann s Disease Juvenile Vertebral Osteochondritis

There is extensive nuclear prolapse into the spongiosa through the cartilage plates The prolapse occurs at the sites of the gaps in the bone at which chondrification had occurred and is due to points of weakness caused by degenerated vessels It is believed that growth in height of the vertebral body is exclusively a function of the cartilage plate This plate lies centrally to the ring epiphysis and beneath the edge of the vertebra Nuclear prolapse in juvenile kypohsis occurs along the vertebral bodies in the lower thoracic and upper lumbar regions The wedging of the vertebral bodies in Scheuermann s disease is secondary to improper mechanical function of the narrowed intervertebral discs and is the cause of the associated kyphotic deformity The fragmentation of the anterior portion of the ring epiphysis results from abnormal pressure relationships and the shearing stress on the anterior annulus fibers and the unfused ring epiphysis

with the blood supply are the apparent causative factors. Roentgenographically, there is flattening and irregularity of the distal end of the involved metatarsal bone(s), a crescentic shape may be present in some instances. The end of the bone is widened, increased in density and may show small, rounded, punched out areas of increased radiance. The proximal end of the adjacent phalanx is usually broadened and flattened and the metatarso phalangeal joint is widened. Loose bones may occur in the involved region. Subchondral necrosis occurs with complete obliteration of the articular surface. The joint is preserved.

OSTEOCHONDRITIS OF THE CALCANEUS

Aseptic necrosis of the calcaneus is manifested by flattening, narrowing and mottling of the apophysis of the calcaneus. A traumatism may call attention to the condition or initiate studies which reveal the lesion. The trauma is not considered the causative factor. On roentgen examination, the apophysis of the calcaneus reveals mottled areas of increased density with irregularity of outline, flattening and narrowing. The adjacent portion of the calcaneus is roughened. There is usually atrophy of the bones of the foot and ankle. The process is self-limited and spontaneous recovery is the rule.

JUVENILE VERTEBRAL OSTEOCHONDRITIS (SCHEUERMANN'S DISEASE)

Scheuermann's disease is also known as adolescent kyphosis or osteochondritis vertebralis juvenilis. The lesion affects the thoracic spine and usually involves several vertebræ. It is characterized by pain and kyphosis in the dorsolumbar region. There may be tenderness over the affected area. Muscle spasm is slight or absent and there is usually no wasting of the muscles. Movements of the spine are limited, particularly flexion. The symptoms subside rapidly with bed rest and immobilization of the spine in a plaster jacket or brace.

Roentgen Manifestations. In patients in whom the disease occurs before the secondary centers of ossification in the epiphyseal rings have appeared, the only demonstrable abnormality is wedging of the affected vertebral bodies. After ossification of the rings the associated features of osteochondritis develop with irregularity of the upper and lower epiphyseal rings, Schmorl's nodes, and sclerosis of the adjacent vertebral surfaces. The intervertebral discs between the affected vertebræ may be narrowed and this causes confusion with tuberculosis. However, there is no bone destruction, rarefaction, or abscess formation in Scheuermann's disease.

Pathogenesis. Adolescent kyphosis is primarily a lesion of the intervertebral discs and is associated with herniation of disc material into the vertebral bodies, the so called Schmorl's nodules. The herniations cause increased *pressure between the adjacent vertebræ, the effect being more* marked along the anterior aspects of the vertebræ. Roentgen changes are frequently demonstrable in the absence of symptoms. The onset of the disease may be associated with vigorous exercise or heavy work. The condition is not due to infection. Scheuermann noted that it was similar

evaluation. Fracture of a vertebra, particularly Kummell's disease, juvenile vertebral osteochondritis, osteomyelitis, metastatic neoplasm, eosinophilic granuloma, xanthomatosis and Gaucher's disease must also be taken into consideration in differential diagnosis.

OSTEOCHONDRITIS DISSECANS

Osteochondritis dissecans is predominantly a disease of young adults and is more frequent in males. The condition may be bilateral although in some cases the symptoms may be present only on one side. While most common in the knee, it has also been observed in other joints and may affect non-weight bearing joints. The most frequent site is the medial femoral condyle. The typical case is characterized by separation of a small rounded, oval or triangular fragment of bone in the subarticular region. The fragment is surrounded by a narrow translucent zone. In the case of the femur, the involved area is usually on the lateral side of the medial condyle near the intercondyloid notch. The zone of rarefaction may fill in with normal bone and all traces of the condition disappear. A narrow band of translucency at the lateral aspect of the medial femoral condyle frequently produces a picture which closely suggests the presence of osteochondritis dissecans. This is a normal phenomenon and is due to the variation in the width and shape of the lower medial border of the inner femoral condyle as it extends upward and laterally. The medial aspect of the intercondyloid notch is thin in some individuals, the borders of the triangle being sharp and well defined. An important differential point is that the anatomical triangle is superimposed on the medial tibial spine while osteochondritis dissecans occurs medial to this area.

Osteochondritis dissecans constitutes an aseptic necrosis of a segment of subchondral bone and results in the formation of an osseocartilaginous sequestrum in the joint. The pathogenesis has not been established. The condition is not infectious in origin. Direct trauma with partial or complete separation of a fragment of bone and cartilage is known to be the causative factor in some cases. However, there are many in which a careful history fails to elicit a history of trauma. There is apparently a familial tendency. As the site of the osteochondritis in the knee and other joints is well protected in all positions of the joint and the roentgen appearance is not that of a fissure fracture, it appears unlikely that direct trauma is the cause. The changes are very similar to those which occur in Legg Perthes and Kienbock's disease. The length of time required to produce the changes in the bone is not known.

Pathology. Layner has described the changes in the bone in a specimen removed at operation. The lesion involved the medial femoral condyle. There was flattening of the affected area and fragmentation of the sequestrum. The cartilage at the margins of the defect was incised and the fragment separated easily. The sequestrum was in a bony recess and was maintained in position by intact articular cartilage and synovial membrane. The lesion may be demarcated by a layer of thin fibrous tissue. When freely movable in the articular space, the fragment tends to become rounded and increases in size by the accretion of cartilage. It may be divided into several pieces. Histopathologic examination of the osteochondritic focus reveals that the center of the affected area is formed by necrotic bone with absence of lamellar substance or an irregular mass of

The mode of longitudinal growth of the human vertebral bodies is still a subject of controversy Beck and Copel were of the opinion that the body of the human vertebra grows in the same manner as the diaphysis of the long bones there being true proximal and distal epiphyseal plates The term "epiphyseal ring is a misnomer and should be discarded as the ring takes no active part in the growth of the vertebral body and is actually a cartilaginous structure which is ossified separately The vertebral ring lies outside the metaphyseal area and fuses with the body when longitudinal growth has been completed It, therefore, appears that the vertebral rings act in the same manner as the apophyses elsewhere in the body and are merely peripheral structures Knutson has studied the growth of the vertebral bodies The irregularities of the bodies characteristic of Scheuermann's disease served as indicators from which measurements were made He showed that anteroposterior growth takes place exclusively in an anterior direction there being no demonstrable increment of growth from the posterior surface The disturbance in Scheuermann s disease appears to be in the vertical growth of the vertebral body This leads to a sagittal wedge formation when the anterior segments of the growth zone are predominantly involved, while in other instances there is a frontal wedge shape The affected vertebra may retain the normal rectangular shape but become smaller than the adjoining normal vertebrae The wedging of Scheuermann s disease is differentiated from compression fracture by the fact that, in the former condition, the sagittal depth of the involved body is greater than that of the adjacent normal vertebra

VERTEBRAL OSTEOCHONDRITIS CALVE S DISEASE VERTEBRA PLANA OSTEONECROTICA

Calvé, in 1924 made the first report of the disease which has been known by his name It is relatively rare He considered five manifestations essential to establish the diagnosis a single vertebra is usually affected although in rare instances two or more vertebrae may be involved the adjacent intervertebral discs are intact cartilaginous new bone formation occupies the space between the discs and the very thin central osseous lamina, the opacity is more marked than that of normal bone, indicating that there is increased osseous density and evidence of regeneration of the osseous nucleus *It occurs under the age of ten years and affects* only the bodies of the vertebræ The disease is similar to coxa plana or Legg Perthes disease of the hip and is not an inflammatory process although it may present many of the clinical aspects of Pott s disease It affects the lower portion of the dorsal spine as a rule only very few instances having been reported with localization of the disease in the lumbar region The affected vertebral body is markedly narrowed and shows increased density The adjacent intervertebral discs remain intact although in some cases there is slight widening of the intervertebral space Regeneration occurs late usually after the third year of the disease Early diagnosis and prompt treatment offer the best hope for recovery

Differential Diagnosis Pott s disease gives rise to very strikingly similar manifestations particularly in the initial stages The roentgen changes alone are frequently not conclusive the history clinical manifestations laboratory findings and serial roentgen studies being necessary for final

evaluation. Fracture of a vertebra, particularly Kümmell's disease, juvenile vertebral osteochondritis, osteomyelitis, metastatic neoplasm, eosinophilic granuloma, xanthomatosis, and Gaucher's disease must also be taken into consideration in differential diagnosis.

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ossifying cartilage with foci of necrosis at the periphery A thick layer of hyaline cartilage is present on the articular surface and a thin layer of fibrocartilage on the opposite aspect

Incidence and Sites of Occurrence The condition is most common in the medial femoral condyle Other frequent sites are the lateral femoral condyle, the head of the femur, the *capitulum* the *supratrochlear septum* of the humerus the head of the radius, the superomedial and superolateral aspect of the talus, the patella at the posterior articulating surface of the lower pole, the first metatarsal bone, usually in the head, and the phalanges, as a rule at the proximal end

Symptomatology The clinical manifestations vary with the site of involvement and the stage of the disease In some instances, there is a well demarcated, undetached sequestrum as an incidental finding in the roentgen examination after a traumatism or when a normal extremity has been examined for comparison These comprise slumbering or latent cases Prior to the detachment of the fragment, the usual complaints are pain and stiffness after walking, prolonged standing, or unusual physical exercise There may be swelling of the joint After the sequestrum has become detached the history is that of repeated sudden locking of the joint similar to that in loose joint bodies from other causes In some instances the patient learns to disengage the fragment and is able to carry on normal duties A sequestrum which has become adherent to the joint capsule results in disappearance of the locking There may be limitation of motion of the joint On physical examination, the affected joint may present no abnormalities or show only slight swelling and limitation of motion Atrophy and wasting of the muscles may occur In rare instances the loose bodies may be palpable

Roentgen Manifestations The roentgen appearance is characteristic and permits of a definite diagnosis In the typical case, the articular surface of the involved bone shows a concave defect measuring about 1 0 cm in diameter The margins are usually smooth but may be slightly wavy In rare instances the margins are undercut irregularly Within the concavity but distinctly separated from its margins there is an ovoid, dense sharply marginated homogenous bony fragment The sequestrum may be divided into several portions In some instances, the area of radiance appears disproportionately large in relation to the sequestrum within it This is probably due to separation of one or more fragments The concavity may be shallow with long, sloping margins probably indicative of an attempt at healing In some instances, the loose fragment is larger than the site of origin This indicates that in its separated state it has become larger by accretion of bone In long standing cases there may be marked osteophyte formation which obscures the bony defect

Differential Diagnosis The syndrome must be differentiated roentgenographically from other conditions in which loose bodies appear in the joint and clinically from diseases accompanied by pain swelling, and locking of the joint The principal lesions which should be considered in differential diagnosis are traumatic loose bodies osteochondromatosis detached osteophytes, neuroarthropathies and tuberculosis Loose bodies in the joint space may result from a fracture with displacement of a bony fragment There is a history of trauma to the region of the joint and roentgen examination demonstrates a fracture The presence of blood, the associated fracture and the presence of a fragment of bone establish the diagnosis in early cases If operation is delayed, the blood is absorbed or

organized and there is regression of the hemarthrosis with degeneration of the separated fragments In this instance the lesion may not be distinguishable from osteochondritis dissecans In synovial osteochondromatosis the condition is the result of hypertrophy of the synovial villi or benign tumors of the synovial membrane Loose bodies are present The bodies are multiple and as many as 100 or more may fill and distend the joint space They tend to be disc like and are dense at the center and periphery with mottled calcification between the central and peripheral portions The number of loose bodies the mottled calcification of the fragments, and the absence of a bony defect distinguish this condition from osteochondritis dissecans Osteophytes may become separated from the margins of the bones in degenerative osteoarthritis and cause locking of the joint as with other joint mice This condition occurs in patients of a much older age group and is associated with polyarticular involvement There is roentgen evidence of irregular eburnation of the articular surfaces of the bones accompanied by marginal lipping or spurring The characteristic bone defect is absent In joints affected secondary to diseases of the peripheral sensory innervation there frequently develop changes which simulate loose bodies This occurs most commonly in syringomyelia tabes and leprosy Because of the loss of sensation and repeated trauma there is eburnation osteophyte formation and fragmentation of the articular cortex with the subsequent formation of loose bodies and narrowing of the joint space These conditions occur in the older age group and the roentgen features are characteristic With a ruptured meniscus there is a typical clinical history and the roentgen manifestations of osteochondritis dissecans are absent One of the most important conditions to differentiate from osteochondritis dissecans is caisson disease In this condition there are multiple infarcts of the long bones usually in the diaphysis although they may also occur in the epiphysis When the epiphysis is involved there develop varying amounts of collapse of the weight bearing portions invasion and replacement by new bone, and calcification of the nonsubstituted portion

Treatment The treatment may be conservative or surgical Cases which are asymptomatic and in which the sequestrum is not detached require no treatment In some instances there is spontaneous healing with disappearance of the necrotic changes Physiotherapy may be helpful Those which do not respond to conservative measures require surgical intervention Removal of the loose bodies is the only treatment which is satisfactory Long standing cases with multiple fragments do not always attain a satisfactory result because of associated arthritis and synovitis

Osteochondritis Dissecans of the Shoulder

Osteochondritis dissecans involving the shoulder is extremely rare There is a relatively high incidence of the disease in certain families and also a high frequency of bilateral involvement On roentgen examination there is a loose body in the joint with evidence of a niche in the articular surface There is little or no sclerosis of the adjacent bone In some instances the loose body remains in contact with the articular surface and is demarcated by an area of increased radiance The bony surfaces of the fragments become covered with dense connective tissue and cartilage A similar change takes place within the defect at the site of origin of the fragments There is usually an effusion of fluid and villous hypertrophy

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Incidence and Sites of Occurrence The condition is most common in the medial femoral condyle Other frequent sites are the lateral femoral condyle, the head of the femur, the capitulum, the supratrochlear septum of the humerus, the head of the radius, the superomedial and superolateral aspect of the talus, the patella at the posterior articulating surface of the lower pole, the first metatarsal bone, usually in the head, and the phalanges, as a rule at the proximal end

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enlargement is not proportional to the alteration in contour of the capitulum. An important manifestation is epiphyseal prematurity, there frequently being premature fusion of the lower humeral and upper radial epiphyses. The union begins on the lateral aspect and proceeds medially, which is not the usual sequence. It is essential to distinguish this change from premature fusion of the epiphysis which occurs after crushing injuries of the cartilaginous plate. The growth stimulation of an injury may continue to act over a period of many years and cause enlargement of the bones but the pattern of the fusion is not disturbed. Bilateral involvement is not infrequent. Cases have been reported in which both elbows and both knees and both knees and one elbow have been affected. No hereditary or familial factors have been found to be constantly present, although there have been instances of multiple cases in the same family. There is a history of injury in approximately 50 per cent of the cases. The disease occurs as a rule in males. There is a tendency for multiple loose bodies to be formed at different times in the course of the disease. The onset is usually in adolescence. There is a predilection for the right side. Osteochondral fractures may lead to loose bodies. Since these are intra-articular, there is usually an effusion. It is believed that the baseball or tennis elbow is not related to osteochondritis dissecans. An important point in this regard is that in a group of 528 pneumatic drill workers suffering from occupational disability of the elbow, only 7 showed osteochondritis dissecans. The sex and age distribution are similar to those in Legg-Perthes disease. The predilection for the right elbow and the convex and articular surface is best explained on the basis of a local anomaly.

Osteochondritis Dissecans of the Knee

In the knee the lesion is visualized in both the anteroposterior and lateral projections although it may be demonstrated to better advantage in one projection than in the other. The so-called posteroanterior 'tunnel view' with the knee partially flexed is of value to demonstrate the intercondyloid region of the femur and may reveal the changes when no other projection proves satisfactory. It is obtained by having the patient lie prone with the knee flexed at 45 degrees, the central ray being projected through the knee joint tangential to the superior surface of the condyles of the tibia. With this view, loose bodies which might ordinarily be obscured are projected into the intercondylar space. In 50 per cent of the cases the defect lies in the region of the central portion of the articulating surface of the medial condyle. Involvement of the external condyle of the femur is rare. The entire condyle may be flattened and the flattening may extend to the lateral margin of the bone in a step-ladder fashion.

In the patella the disease is demonstrated as a defect in the otherwise smooth, slightly concave posterior surface of the bone. The lower pole is most commonly involved. The sequestrum may be small and cast a very faint shadow, hence is easily overlooked. The tangential view, either inferosuperior or superoinferior, may be of great aid in demonstrating the lesion. In making this roentgenogram, the patient is prone with the knee fully flexed and the central ray is directed perpendicularly between the patella and the femur. It may also be made with the patient supine and the knee slightly flexed, the ray being directed toward the feet.

Osteochondritis Dissecans of the Elbow

Osteochondritis of the elbow is relatively common Clinically, the condition is manifested by limitation of extension and palpable enlargement of the radial head The absence of marked synovial swelling serves to differentiate the lesion from chronic infectious diseases The typical appearance of a small fragment of subchondral bone surrounded by a zone of rarefaction which is pathognomonic of this lesion is rare in the elbow The earliest roentgen manifestation is best demonstrated in the anteroposterior projection as a patchy area of rarefaction with sharply defined borders The process usually affects the convexity of the capitulum In some instances there is an irregular, cyst-like area which is best described as a



Fig 285 Osteochondritis Dissecans of the Humerus There is a small rounded fragment of bone surrounded by a narrow zone of increased radiance at the articular surface of the capitulum

series of signet rings of different sizes lying within each other There may be an appearance suggestive of the formation of multiple small fragments of bone *Rarely there is a solitary large area of rarefaction without a* sequestrum In this instance the condition may be confused with tuberculosis There may be a loose fragment of bone adjacent to the capitulum Irregularity of outline and cyst formation in the capitulum may also occur In the majority of cases there are loose bodies indicative of the presence of a previous osteochondritis The lesion may present eburnated irregular margins Secondary osteoarthritis frequently develops and has been considered the cause rather than the result of the loose bodies In long standing cases the sigmoid cavity of the ulna may be large and deep and present angular margins rather than the normal smooth curve

The head of the radius frequently shows enlargement The enlargement may occur before or after closure of the epiphysis The degree of

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series of signet rings of different sizes lying within each other There may be an appearance suggestive of the formation of multiple small fragments of bone Rarely there is a solitary, large area of rarefaction without a sequestrum In this instance the condition may be confused with tuberculosis There may be a loose fragment of bone adjacent to the capitulum Irregularity of outline and cyst formation in the capitulum may also occur In the majority of cases there are loose bodies indicative of the presence of a previous osteochondritis The lesion may present eburnated irregular margins Secondary osteoarthritis frequently develops and has been considered the cause rather than the result of the loose bodies In long standing cases the sigmoid cavity of the ulna may be large and deep and present angular margins rather than the normal smooth curve

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CONDENSING OSTEITIS OF THE ILIUM

The term condensing osteitis of the ilium is used to describe a disturbance of the normal architecture of the ilium characterized by an area of osteosclerosis in the region adjacent to the sacro iliac joint It is a condition which is recognizable only roentgenographically Roentgen studies show increased density in the auricular portion of the ilium with absence of corresponding changes in the sacro iliac joint or the contiguous portion of the sacrum Berent in 1932 suggested that it might be a complication of pregnancy Rendich and Shapiro recorded a case in 1936 In 1944 Hare and Haggart reported a series of female patients with this condition some of whom were nulliparous and stated that the condition might represent a disturbance of the circulation In several of the patients there was



Fig 288 Bilateral Condensing Osteitis of the Ilium There is a narrow band of increased density in the medial aspect of the ilium adjacent to the sacro iliac joint on each side The sacro-iliac joints are not involved

a history of trauma in some instances related to childbirth, which may be considered a possible cause for alteration in the blood supply in the inferior portion of the ilium in the region adjacent to the sacro iliac joint

The sacro iliac joint is an amphiarthrosis the auricular surfaces of the sacrum and ilium being separated by thin plates of cartilage In the young adult these cartilaginous plates are intimately blended with each other During later years, they become separated by fluid with the formation of a joint space thus converting the articulation into a diarthrosis The motion involved in this joint is minimal the sacrum tending to rotate forward about a fulcrum situated in the vicinity of the second sacral segment The sacrum does not actually form a keystone in the posterior arch of the pelvis Its articular surfaces are more widely separated anteriorly than posteriorly and for this reason the bone tends to be displaced forward into the pelvis with the patient in the weight-bearing position The interosseous and posterior sacro iliac ligaments resist this by becoming taut thus forcing the iliums more closely together An additional factor which interferes with the stability of the articulation develops



Fig 286 Osteochondritis Dissecans of the Femur There is an area of increased radiance and irregularity of outline involving the articular surface of the medial condyle of the femur A small island of bone is visualized within this area The patient complained of pain and limitation of motion of many months duration Operation confirmed the roentgen diagnosis of osteochondritis dissecans



Fig 287 Osteochondritis Dissecans of the Knee There is irregularity of outline and increased radiance in the lower end of the femur and a small fragment of bone lying within the knee joint

occurs as a rule in the female There is an elevated sedimentation rate in arthritis Roentgenographically, there is narrowing or obliteration of the joint space in Marie Strümpell arthritis with marked osteoporosis of the bones Other conditions which may cause confusion in diagnosis are sacro iliac epiphysitis and asymmetry of the lumbosacral facets Paget s disease and degenerative arthritis are excluded on the basis of the age of the patient



Fig 289



Fig 290

Fig 289 Bone Infarct There is an irregular area of increased density within the medullary cavity of the upper third of the humerus the characteristic manifestation of a bone infarct

Fig 290 Bone Infarct There is an irregular area of increased density in the upper end of the left femur which represents an infarct

BONE INFARCTS AND ASEPTIC NECROSIS IN CAISSON AND OTHER WORKERS

Roentgen examination of individuals who have worked under compressed air in some instances reveals single or multiple infarcts in the shafts of long bones The infarcts may occur in association with areas of aseptic necrosis in the joints The disease caused by too sudden release of the pressure after working under compressed air is variously described as caisson disease aero embolism or the bends Aero embolism has been reported in aviators and deep sea divers but bone and joint changes have not occurred in these individuals Similar changes have been described in the bones and joints of individuals who have never worked under compressed air Taylor discusses a group of 54 patients with aseptic necrosis and bone infarcts Thirteen were women Of the 41 men 12 had a history of working under compressed air while the remainder had no occupational history Of the 13 patients who had worked under com

during pregnancy, as relaxation of the sacro iliac ligaments and other pelvic ligaments occurs The auricular portion of the ilium resists the thrust of the sacrum as the latter rotates It is believed that the pressure exerted upon the para articular surface of the ilium is the cause of the condensation which is noted roentgenographically

Pathological Findings Rendich and Shapiro described the micro scopic appearance of an iliac bone removed from a patient with condensing osteitis They noted that the lacunæ were obliterated by extensive con densation of the osseous tissues Shipp and Haggart state that their experience has been at variance with these findings and indicate that the discrepancies may represent different phases of the same process They in terpreted the changes as representing a low grade, nonspecific ischemic process

Symptoms Chronic low back pain is the chief complaint The pain differs from that in sacro iliac arthritis or radicular irritation and radiates to one or both buttocks, never along the sciatic distribution There is no aggravation of symptoms as a result of coughing or straining, although the symptoms may be caused or increased by activity Relief is obtained with rest The symptoms are seldom severe enough to be incapacitating, but usually increase slowly in severity and tend to become continuous In many instances, the onset of the pain is during the final trimester of pregnancy or immediately after delivery, with recurrence or exaggeration during a subsequent pregnancy In other patients symptoms appear to follow rapid gain in weight, excessive physical fatigue or other debili tating factors History of severe trauma to the back is seldom present In most of the reported cases the patients have been white women be tween the ages of nineteen and forty-four, the average being thirty-two years The symptoms are present for an average of four years prior to the establishment of the diagnosis the period ranging from four months to eighteen years However the disease has been described in young, unmarried females and also in males

Physical Examination The physical examination is usually negative except for an increase in the lumbar lordosis In severe cases there may be spasm of the erector muscles Tests for sacro iliac joint disease are negative A common finding is inadequate musculature which may be associated with moderate obesity The laboratory findings are not remarkable

Roentgen Findings The routine roentgen study of the lumbosacral spine in the anteroposterior and lateral projections is not satisfactory for visualization of the sacro iliac joints It is essential that a flat sacral projection be utilized The affected portion is behind the anterior margin of the joint so that condensation in this region may convey a false im pression of involvement of the sacro iliac joint Stereoscopic studies show that both sacro iliac joints and the adjacent sacrum are normal in appear ance There is dense sclerosis in the auricular portion and sometimes in the portion of the ilium adjacent to the sacro iliac joint In this area the normal bony trabeculæ are obliterated There is usually no sharp line of definition between normal and abnormal bone It is important to stress the fact that there is no correlation between the roentgen manifestations and the severity of the symptoms

Differential Diagnosis Condensing osteitis is most often confused with sacro iliac arthritis particularly of the Marie Strumpell type This con dition occurs more commonly in the male while osteitis condensans ilii

been suggested that nutritional disturbances such as occur in scurvy and rickets might be responsible. The fact that the islands are most common in the region adjacent to the joints and are usually unilateral and solitary indicates that trauma may be an important factor. In 24 per cent of Steele's cases the patients exhibited some form of arthritis. Some authors have attempted to explain chronic arthritis on the basis of primary aseptic necrosis of the bone resulting from an injury to the blood supply at the junction of the bone and the cartilaginous lining of the bone. If it is assumed that the insular calcific deposits are bone infarctions or the sites of organized aseptic necrosis, it would be expected that they would occur with a high degree of frequency in arthritis. This is partly borne out in Steele's patients. In 27 per cent of the cases with bone islands there occurred associated blood vessel abnormalities. If the calcified medullary defects are believed to be small asymptomatic organized infarcts, a high incidence in individuals with vascular pathological changes appears logical. In some of the previously reported cases arteriosclerosis was the outstanding finding. Chandler has demonstrated bone infarcts in the head of the femur after obliteration of the nutrient arteries. It has been shown experimentally that the epiphyseal arteries undergo transformation from transepiphyseal structures in childhood to end arteries in adult life. These terminal arteries are particularly prone to blockage and subsequent anemic infarction of the tissue receiving blood supply from these arteries is to be expected. It is important in this connection to stress that 73 per cent of the defects occurred in the epiphyseal line area where a bland infarct might have occurred in an end artery. The studies of Langer in 1875 demonstrated conclusively that the bones contain end arteries. Changes which may develop in the carpal, tarsal and metatarsal bones and the articular end of the femur subsequent to vascular occlusion have received considerable attention. Reference to infarcts of the shafts of the long bones are very infrequent. The reason probably lies in the fact that these lesions are, as a rule, silent in nature. Roentgen demonstration of this lesion is usually accidental and biopsy studies are not warranted. Similarly, during autopsy the pathologist only rarely has occasion to examine the long bones; hence infarcts are not found at postmortem unless a preliminary roentgenographic study has been made and special search instituted.

Roentgen Manifestations. The roentgen manifestations of infarcts involving the diaphysis are characteristic. There is a well defined, often symmetrical area of mottled density, usually in the medullary portion of the diaphysis. In some instances the changes extend into the epiphysis. Involvement of the non-articular cortex is rare. The area of density varies from a mere fleck to several centimeters in diameter. The larger lesions may be demarcated from the adjacent bone by a narrow band of marked calcification. The lesion may be manifested as a cystic area with dense calcification about the margins of the area of increased radiance. The affected area may become completely calcified and present a homogeneous dense, non-reticulated appearance. The infarcts may be single or multiple, vary widely in distribution and may occur in practically any portion of the bone. In rare instances the lesions are bilaterally symmetrical. Though histologic proof is lacking in many instances, it is possible to establish the diagnosis with a high degree of certainty by roentgen methods.

Differential Diagnosis. In osteopoikilosis there are asymptomatic scattered areas of density in the bone. These are widely distributed

pressed air, some had been subjected to sudden changes of pressure and had developed symptoms of aero embolism or bends, in those who had not been subjected to sudden changes in pressure some had mild symptoms of decompression sickness while others were asymptomatic Lesions of the bones and joints do not develop immediately after decompression The bone lesions are generally asymptomatic Arthritic changes resembling those of chronic hypertrophic osteoarthritis constitute the usual associated manifestations in the joints The lesions in the non occupational group were the same as those in the occupational group and could not be differentiated by roentgen methods One of the cases of aseptic necrosis was in a patient who had sickle cell anemia and a history of polyarthritis The etiology of the non occupational cases is not known The bone lesions are generally multiple and often are bilateral

Calcified Islands in Medullary Bone Infarcts Islands of calcification in medullary bone are relatively common The nature of these shadows is not clearly understood They are usually asymptomatic unilateral, irregular, insular areas of increased density in the bones Most roentgenologists classify them as calcified medullary defects of no pathological or clinical significance The islands occur most commonly in the ends of the long bones and are usually situated centrally in the cancellous portions Bony trabeculae are absent in the region of the islands As a rule they are oval in shape although many are amorphous irregularly demarcated areas of calcific density The cortical bone in this region is not deformed and there is no evidence of expansile changes In none of the previously reported cases have there been any symptoms which could be referred to the area of the lesion Most commonly the change is found in the course of a roentgen study for arthritis or fracture It has not been possible in any case to prove that the medullary islands are responsible for the patient's complaints

In a study by Steele a review was made of 6 000 roentgenograms and 120 lesions of this type were found The most common site was the femoral neck there being 25 per cent in this region The next most frequent region of involvement was the condyle of the femur, which comprised 17 5 per cent In the wing of the ilium there were 13 3 per cent, the proximal portion of the tibia 12 5 per cent and the head of the humerus 7 5 per cent Less common locations were the shafts of the femurs, the calcaneus the ischium the head of the fibula the metacarpals the condyle of the humerus the patella and the vertebrae In the femoral neck 11 per cent of all the cases showed calcified medullary defects and of the femoral condyles examined 9 6 per cent showed these lesions None were found in the ribs the scapula the clavicle the thoracic vertebrae or 3,800 chest plates The distribution as to the right and left sides was approximately equal The calcified islands have been noted in patients from the teens to the eighth and ninth decade of life No definite correlation is possible between the patient s antecedent or co existent illness and the presence of the medullary defects In rare instances the defects are multiple but as a rule they are unilateral and solitary In none of the cases studied was there a history of submarine duty deep sea diving or caisson disease In 11 per cent of the cases a two year follow up study was made and no change in the defects was demonstrable

Etiology No satisfactory explanation as to the etiology of the defects has been advanced Developmental anomalies may be involved faulty ossification of cartilage being considered the important factor It has

Chapter

8

Neoplasms of Bone Tumors of the Skeleton

Introduction A tumor is defined as an independent or autonomous overgrowth of tissue which does not serve a useful purpose By use of the term independent or autonomous the tumor is characterized as being outside the genetic pattern of the individual and indicates also that it is out of balance with the other tissues of the body The tumor fulfills no useful purpose, in contradistinction to proliferative tissue which serves as a defense mechanism in the body, as for example, granulation tissue which develops during inflammation or regeneration Tumors may be either benign or malignant The benign tumors are characterized histopathologically by marked differentiation of their elements and in most instances resemble the mother tissue They show a moderate rate of growth and enlarge by expansion with resultant dislocation and compression of the neighboring tissues and organs Although usually solitary they may be multiple They do not produce metastatic lesions While a benign tumor does not kill its host directly it may become fatal by compressing or displacing vital organs such as the brain A malignant tumor is generally characterized by cells of lower differentiation than those of the mother tissue It grows slowly or rapidly and invades the surrounding tissues and organs Because of its tendency to invade the blood or lymph vessels elements of the tumor may be carried to remote regions of the body giving rise to metastatic lesions The histologic differential diagnosis between benign and malignant tumors may be extremely difficult The decisive or determining factor in establishing the fact that the tumor is malignant in nature is the demonstration of invasive growth of the tumor cells The level of differentiation of tumor cells as compared to that of the mother tissue helps to determine the degree of malignancy of the tumor The lower the differentiation of the tumor cells the more highly malignant the neoplasm

Roentgen examination is of the greatest value in establishing the presence of bone tumors and frequently makes it possible to determine the exact nature of the neoplasm Closely similar manifestations may occur in lesions due to trauma inflammatory processes, developmental anomalies metabolic disturbances and other neoplasms Once the presence of a lesion has been demonstrated roentgenographically careful correlation of the clinical manifestations and the roentgen changes is essential The decision as to whether the lesion is neoplastic in character can be made in many instances, and it is usually possible also to determine whether it is benign or malignant Certain neoplasms such as osteogenic sarcoma metastatic carcinoma from the breast or prostate giant cell tumor and others present such characteristic roentgenographic features that an accurate diagnosis can be made

throughout the body The areas of condensation are believed to be hypertrophied trabeculae of the spongiosa and are most common in the metaphysis or the epiphysis In osteoid osteoma, the lesion may involve any bone and has been described in the skull the scapula, and the clavicle, sites in which the medullary calcified islands have not been found Osteoid osteoma is associated with bone pain of several months duration and is usually seen in the younger age groups There is localized swelling and tenderness, manifestations which are not present in the insular defects Osteoid osteoma begins as a rarefied area in the bone and progresses to dense calcification There is usually a radiolucent center with a characteristic nidus Osteogenic sarcoma, particularly of the sclerosing variety, is associated with severe pain, worse at night The roentgen finding is that of a poorly defined bone lesion with cortical invasion in contradistinction to the central, well defined appearance of the calcified islands Metastatic neoplasms are easily ruled out In these lesions, the defects are disseminated and multiple Metastases from prostatic carcinoma are most commonly found in the pelvis and the lumbar vertebrae While they are frequently osteoblastic they may be osteolytic which aids in the differentiation from calcified medullary defects In chondrodysplasia the defects are due to faulty ossification of cartilage The hereditary form of dyschondroplasia is in many instances characterized by roughly symmetrical masses of cartilage which originate in cortical bone Bone infarcts are usually found at the ends of the long bones and this is a common site for the calcified defect There is usually an area of density which surrounds the infarct and appears to be due to re ossification and repair Also in infarcts there is usually central translucence It has been stated by some authors that caisson disease is manifested by bone infarcts the compressed gas being the etiologic agent The description of the lesions in the reported cases is identical with that of bone infarct The lesions are extensive frequently bilateral and multiple and an occupational history is usually elicited

growing tumor may cause resorption on the surfaces of the trabeculae facing the tumor and compensatory formation of new bone on the opposite surface of the trabeculae. The resorption and formation of new bone may result in gradual changes in the bone, bending, expansion or swelling of the bone comprising manifestations of these changes. Destruction of bone by tumor may be rapid. Compensatory formation of new bone with consequent strengthening of the weakened bone is also rapid. In most instances, the repair is accomplished by the development of osteophytes, a peculiar type of spongy bone which may develop in many conditions. Osteophytes are not confined to any specific skeletal disease. Bone shows practically no resistance to the growth of a tumor and is unable to check its expansion, connective tissue being much more resistant to the pressure exerted by a proliferating neoplasm. The changes in the periosteum during compensatory bone formation comprise an elevation of the periosteum from its normal position by the apposition of new bone. This is not a passive mechanism as the periosteum grows with the growing bone and constitutes an active, co-ordinated biologic process. In the shaft of a long bone at the point of junction of the newly formed bone and the premorbid bone, the new bone projects from the old in a wedge-shaped layer. The lipping of the periosteum is frequently clearly demonstrable by roentgen methods. This is termed *Codman's triangle* and comprises an important factor in the diagnosis of certain tumors of bone. The newly formed bone may exceed the tumor in extent both proximally and distally. The spur or osteophyte is visible even after the tumor has caused resorption of the newly formed bone with which it is in contact.

Cartilage in the epiphyseal plate or the articular cartilage is very resistant to the encroachment of a tumor. A tumor is frequently separated from the cavity of a joint by articular cartilage and extends into the joint only if this cartilage is partially destroyed mechanically after it has lost the support of the underlying bone. Uncalcified cartilage is almost immune to resorption. Calcification of cartilage is dependent upon preceding degenerative changes. Normal, vital, noncalcifying cartilage persists in spite of the progress of the tumor. If the destruction of bone proceeds at a faster rate than the compensatory formation of new bone, pathologic fractures may develop. Conversely, when the compensatory formation of new bone exceeds the destruction of bone, sclerosis ensues. Metastases of certain epithelial tumors, particularly carcinoma of the bladder and prostate, tend to induce extensive production of bone. If the cells proliferating from metastases of a prostatic carcinoma invade the marrow spaces of the bone, formation of new bone may be observed on all or most of the bony surface adjacent to the tumor. The ability of the tumor cells to induce osteoblastic bone formation from the connective tissue covering the bone stems from richness of the tumor cells in phosphatase. Transplantation of the mucosa of the urinary bladder may induce the surrounding connective tissue to form bone in certain regions of the body.

CLASSIFICATION OF PRIMARY TUMORS OF THE SKELETON

The classification of the tumors of the skeleton is an extremely difficult problem. The difficulties stem in part from the fact that there is a lack of clear differentiation between bone as tissue and bones as organs. The difficulties are increased by the fact that many tumors of the bones show a protean polymorphism. The tumors originate from more or less differ-

Bone neoplasms usually produce alterations in the densities and outlines of the affected tissues. As in early osteomyelitis and many other lesions, malignant growths may be present in bone for considerable periods of time before changes are demonstrable in the roentgenogram. This is particularly true of metastatic lesions of the vertebra.

The benign tumors of bone are manifested by an aberration of normal growth in which the inhibitory factor which normally controls the architecture of the tissues is partially suspended but not entirely lost. The continuity of the contour of the cortex remains intact. The affected portion of the bone shows a change in density. This change may consist of an area of increased radiance or one of increased density, the character and extent of the changes depending on the nature of the tumor. Bone is destroyed by pressure erosion as the result of the growth of the tumor. The margin of the tumor tends to be definite and smooth and is sharply demarcated from the normal bone. As the growth of the tumor continues, a benign neoplasm tends to expand the contour of the bone and alter the continuity of the cortical outline. The neoplasm may invade the contiguous soft tissue structures. The outline of the invasive shadow is clearly demarcated from the adjacent normal soft tissue structures. When the natural control of growth is lost and the tumor appears to grow without restraint, it is a malignant tumor. The malignant nature of the tumor is manifested by dissolution of the cortical contour at one or more points in the bone and in many instances elevation of the periosteum from the underlying cortex in the region of the break in contour. The margins of the foci of destruction in the bone caused by the tumor are irregular and fade gradually into the shadow of the normal bone. These and other similar characteristics serve as important aids in diagnosis.

The Reaction of Bone to the Growth of Tumors. The reaction of the surrounding bone to a bone tumor is dependent on the character of the neoplasm, the rate of growth, and other factors. The bone architecture may not be affected in the early stages. There may be predominant bone *absorption. Except in the presence of very rapid absorption, there is* usually simultaneous osseous reconstruction rather than complete loss of bony architecture. The tumor cells in themselves do not cause absorption of bone. They act by stimulating osteoclasis. The effect of the bone tumor in some instances is chiefly bone deposition. Various types of tumors of widely different histopathologic structure may produce osteoblastic metastases, as for example, carcinoma of intestinal or bladder origin. In many instances it is difficult to distinguish primary from metastatic growths despite careful clinical, roentgen and pathologic evaluation. This is particularly true in Ewing's sarcoma. Osteoblastic reactions to tumors are very similar to the reactions to inflammatory stimuli. A rapid reaction produces a fine network of woven bone in periosteal and/or endosteal tissue. With a slower type of response, lamellar bone is formed on the surface of existing lamellar bone or on a basis of woven bone trabeculae.

Bones react to the growth of tumors in practically the same way whether the neoplasms are benign, malignant, primary or secondary. If *the tumor grows slowly and bone destruction proceeds at a moderate or* slow rate, new bone may be formed in practically the same way as during normal growth. The compact cortical layer is resorbed from within and as a compensatory mechanism, a layer of lamellated, mature bone is laid down on the periosteal surface. In spongy bones, the presence of a slow

which are impractical and indefinite because of overlapping or lack of clear cut differentiation. The Bone Sarcoma Registry of the American College of Surgeons bases its classification of bone sarcomas on histogenesis, the subvarieties being designated by the pathologic anatomy, as this method lends itself to the practical needs of the surgeon, radiologist and pathologist. The myelomas are an exception to the rule and the subvarieties in this group are based on the cell of origin. A modification of the Bone Sarcoma Registry classification will be used as a basis for discussion.

PRIMARY BONE TUMORS

		Malignant	*Benign*
Osteogenic Series			
	Osteogenic sarcoma	Medullary and subperiosteal	Osteoma
		Telangiectatic	Parosteal Osteoma
		Sclerosing	Osteoid Osteoma
		Periosteal	
		Fibrosarcoma	
		Medullary	
		Periosteal	
Chondroma series		Chondrosarcoma	Chondroma
		Myxosarcoma	Periosteal Chondroma
			Epiphyseal Chondroblastoma
			Osteochondroma
			Myxoma
			Non osteogenic Fibroma
Giant Cell Series		Malignant Giant Cell	Benign Giant Cell Tumor
			Solitary Bone Cyst
			Aneurysmal Bone Cyst
Angioma Series		Endothelioma	Hemangioma
		Ewing's Sarcoma	
		Angiosarcoma	
Myeloma Series		Multiple Myeloma	
		Reticulum Cell Sarcoma	

METASTATIC BONE TUMORS

Epithelial Carcinoma
Mesodermal Sarcoma

MISCELLANEOUS

Malignant Lymphoma
Leukemia
Myeloid Metaplasia
Myelosclerosis
Neuroblastoma
Chordoma
Presacral Tumors

BENIGN BONE TUMORS

The benign tumors are usually classified by the type of tissue of which they are principally composed, while in the case of malignant neoplasms the most satisfactory classification is based on the tissue from which the growth appears to arise. The benign neoplasms which originate from the cortex of the bone are osteoma, osteochondroma and chondroma. In

entiated tissue The benign tumors of connective tissue often form cartilage and more frequently bone The undifferentiated connective tissue cells in malignant tumors of the skeleton have many potencies and wide variability of the histologic structure in different regions of the same tumor is a common occurrence There are multiple transitions from one type to the other In consequence, it may prove more satisfactory not to take into consideration the histologic picture rather to subdivide the tumors on the basis of their clinical behavior This is particularly true of the osteogenic tumors because of their development from bone forming tissue The classification of this group is based on the prominence of various structural qualities which, to a certain degree, are common to all osteogenic sarcomas

It is possible to divide the tumors of the skeleton according to the mother tissue only insofar as the tumors of the supporting tissues in the skeleton can be subdivided into fibroma, chondroma and osteoma However, confusion ensues in that an osteoma can be regarded as a tumor of bone tissue when it arises from the secondary ossification of a fibroma and the same is true in the case of a chondroma or an osteochondroma With regard to the malignant growths, it is doubtful whether a fibrosarcoma in its pure form arises from tissues of the skeleton, for although fibrosarcoma may arise from the outer layer of the periosteum, more commonly this tumor originates from the extra skeletal connective tissue such as the fascia and tendon In using the terms "osteogenic tissue and "osteogenic sarcoma' one must stress the fact that bone formation is not an exclusive faculty or function of the connective tissue which forms part of the skeleton, as under suitable conditions and in pathologic cases cartilage or bone may be formed almost anywhere in the connective tissue

A classification of bone tumors according to the mother tissue is used by many authors, particularly with reference to the malignant neoplasms The supporting tissue may be connective tissue cartilage, or bone The benign primary tumors originating in connective tissue is the fibroma which may be ossifying and the myxoma The malignant member of this group is the fibrosarcoma The cartilage gives rise to the chondroma and in the malignant group to the chondrosarcoma The tumors which arise from bone particularly the osteoblasts comprise the osteoma and the osteochondroma with their malignant counterparts termed the sarcomas The tumors which are clasified as arising from accessory tissue are those which originate in the blood vessels the fatty tissue and bone marrow and include (1) the blood forming elements and (2) the reticulum The benign members of this group are the angioma and the lipoma, while the malignant tumors comprise the angiosarcoma and the liposarcoma In the tissues which arise in the bone marrow the blood forming elements give rise to the multiple myeloma while the reticulum cells give rise to Ewing s tumor The secondary tumors are divided into epithelial which give rise to the metastases of carcinoma and the mesodermal, which give rise to the metastases of sarcoma

The classification of bone tumors is still admittedly unsatisfactory It is necessary that the classification be correct from the pathologic point of view and avoid an excessive number of subdivisions yet be sufficiently complete to include all of the neoplasms of whose existence we are aware Many of the subdivisions previously used have been found to be valueless or confusing and are mentioned merely because they are so well established in the literature The best practice is the avoidance of subdivisions

and other irritations Ewing states that spontaneous or traumatic but noninflammatory origin progressive course circumscribed form, active participation of osteoblasts, and derivation from cartilage are prominent features in true osteomas The bony tissue of an osteoma presents solid lamellae and a few Haversian canals It may be spongy and in some instances contain marrow spaces or cavities The dense osteomas arise from membranous bone periosteum or within the bone itself The less dense types originate from cartilage or bone of cartilaginous origin

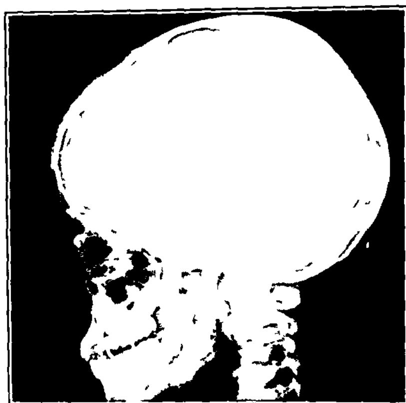


Fig 293 Osteoma of the Frontal Bone There is a large spongy osteoma of the outer aspect of the frontal bone The external surface of the neoplasm is smooth and rounded and appears to blend with the surface of the skull The inner table of the skull is thinned and appears slightly depressed in the region of the osteoma The outer table apparently has preceded the mass The neoplasm appears continuous with the diploe The frontal sinuses are markedly enlarged Roentgenograms which are too dark because of overexposure may not show the osteoma the shadow of the tumor mass being obliterated in the darker portions of the film

Osteoma of the Skull The commonest site of osteomas of the skull is the frontoethmoidal region The middle fossa and petrous pyramids are less frequently involved They usually occur before puberty only rarely beginning in individuals past middle life Males are more frequently affected than females The neoplasms appear to be located in greater numbers at points where the sutures of two bones unite The exostotic form which arises from the outer surface of the bone is more common than the enostotic or those originating from the inner surface If large they may cause headaches and other concomitants of cerebral compression When the orbit is invaded a nonpulsating exophthalmos may develop The intracranial type may in rare cases extend into the lateral ventricle or perforate a sinus The skin overlying exostotic osteomas is movable Osteomas grow very slowly Because of their location or rate of growth,

some instances, it may be difficult or actually impossible to differentiate tumors from bone proliferation due to inflammation, mechanical changes or congenital anomalies, an example of these changes being the bony spurs commonly seen at the tip of the olecranon process and the inferior surface of the calcaneus The benign tumors commonly arising from the medullary portion comprise bone cyst, benign giant cell tumor, enchondroma and fibroma



Fig 291 Fig 292

Fig 291 Osteoma of Tibia There is a large osteoma at the lower end of the diaphysis of the tibia The osteoma has a broad base In the lateral projection there is backward bowing and localized thinning of the fibula at the level of the osteoma

Fig 292 Osteoma of the Scapula There is an osteoma at the anterior aspect of the scapula (arrow) The osteoma was demonstrable only on the lateral view of the scapula

Osteoma

Osteomas are benign bony tumors They are sharply circumscribed arise from the cortex and extend into the adjacent soft tissues The fibrous type occur usually in the flat bones and are most common in the skull orbits facial bones and accessory sinuses, less frequently they are found in the long and short bones Roentgenographically, they form sharply defined irregular or smooth osseous projections from the surface of the bone The density is similar to that of the adjacent bone Osteomas which form within the bone project into the marrow cavity and are known as endosteomas Multiple osteomas may occur as a familial anomaly They are long irregular and spring from the shafts of the bones Not all bony tumors are osteomas for there may be bony overgrowth in association with reparative reactions in certain infections invasive neoplasms

and other irritations Ewing states that spontaneous or traumatic but noninflammatory origin progressive course, circumscribed form active participation of osteoblasts and derivation from cartilage are prominent features in true osteomas The bony tissue of an osteoma presents solid lamellae and a few Haversian canals It may be spongy and in some instances contain marrow spaces or cavities The dense osteomas arise from membranous bone periosteum or within the bone itself The less dense types originate from cartilage or bone of cartilaginous origin

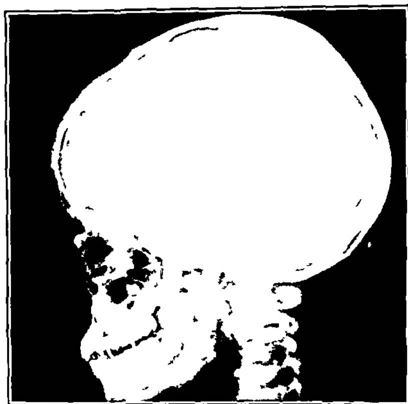


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they may constitute a definite menace to the health of the patient These neoplasms not infrequently continue to increase in size after normal bone growth has ceased Trauma or infection may accelerate their growth Rapid increase in size is an indication for prompt surgical removal as this suggests the possibility of malignant degeneration Osteomas of the skull vary markedly in size and characteristics They

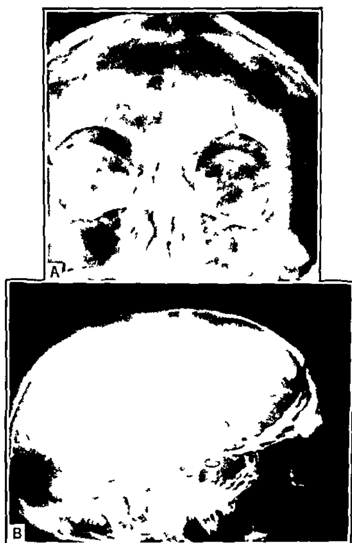


FIG 294 Osteoma of the Frontal Sinus *A* Sagittal projection *B* Lateral view The osteoma is visualized as an area of bony density in the left frontal sinus The margins of the tumor are sharply delineated The density of the osteoma is greater than that of normal bone

may be rounded smooth domelike and of uniform density or present rough irregular surfaces with radiance in the central or basal portion They tend to blend with the surface of the skull There is no ballooning of the tables as a rule although in some instances the appearance is that of diploe which extends from the bone with the outer table of the skull having disappeared or preceding it Osteomas of the cranial surface appear to arise from the inner table of the skull, which may be thickened and slightly depressed The density varies from spongy and radiant to extremely dense The tables of the skull frequently cannot be differentiated from the tumor Spicule formation is rare but does

occur in some instances The frontal sinuses may be enlarged in cases of spongy osteoma of the external cranial surface This occurs particularly if the osteoma encroaches on the glabella In differential diagnosis one must consider meningiomas calcification of the meninges or in a hematoma osteitis frontalis interna and leontiasis ossea or craniosclerosis Meningiomas usually show preservation of the outlines of the tables and deposition of new bone in irregular or regular manner, adjacent to the surface of the tables of the skull Spicule formation is common in meningiomas and there is apt to be marked prominence of the blood vessel grooves in the region of the neoplasm Orbitoethmoid osteoma may extend inward and be confused with a meningioma of the olfactory groove The rate of growth in meningiomas is more rapid than in osteomas Calcification of the meninges is linear triangular or in plaques and is most common in the region of the falx In calcified hematoma there is irregular or mottled calcification which is as a rule, distributed throughout the mass Hyperostosis frontalis interna presents wavy multiple densities along the inner table and because of the age and sex incidence and association with definite clinical symptoms may be differentiated without difficulty Leontiasis ossea occurs principally in the frontal, basal, and facial bones Some observers feel that this is an atypical diffuse osteoma however it is multiple progressive and accompanied by changes in the facies which usually stamp it as a clinical entity quite apart from osteoma The temporal bone may present a small single exostosis of the inner table which closely resembles an osteoma This formation is of the same texture and density as the remainder of the temporal bone and is a normal variation which is not of pathologic significance and must not be confused with a neoplasm

Parosteal Osteoma

Geschickter and Copeland report a new bone tumor entity which they term parosteal osteoma The tumor occurs in the long bones predominantly in the lower femur upper humerus or upper fibula The initial lesion is most frequently a benign proliferation of ossifying fibrous tissue which arises in the region of the periosteum of a long bone A rounded bony mass projects from the shaft of the bone in the region adjacent to the metaphysis The tumor presents no base or pedicle of normal bony growth and there is no overlying cap of cartilage such as occurs in osteoma or osteochondroma The ossifying mass extends by contiguity through the adjacent bone structure at one or more points There is peripheral extension into the soft parts An encapsulating periosteal membrane may be present The tumor undergoes progressive growth Ultimately, there is malignant degeneration The source of the new bone consists of bands of skeletal blastema which have been displaced from the primitive periosteum The points of predilection comprise surfaces of the long bones free of muscular attachments the commonest site being the popliteal fossa The lesion occurs most frequently during early adult or middle life but may also be present in the younger age periods The clinical manifestations comprise swelling pain and a mass which tends to increase progressively in size The tumors are usually present for months or years Trauma does not appear to be of importance in the etiology but may be significant in that the mass is first discovered after an injury There is no increase in the white blood cell count and no elevation of temperature

Physical examination reveals a mass of bony hardness firmly attached to the adjacent bone and extending into the soft tissues Invasion of the muscles is common The periphery of the tumor usually is smooth although in some instances the surface appears irregular and infiltrating The condition is characterized by slowness of growth, discrete margins and normal overlying soft tissues These manifestations indicate that it is benign in character However, the lesions are potentially malignant After surgical removal, there is usually continuous activity and, in many instances, sarcomatous degeneration results Local recurrence is common Pulmonary metastases may occur in some cases five or more years after the removal of the primary lesion

Roentgen Manifestations On roentgen examination, there is a dense irregular, rounded mass of new bone The tumor is separated over most of its circumference from the underlying adjacent bone There may be several discrete, smaller, osseous masses particularly in recurrences after operative removal The base of the lesion is broad and is characterized by widening of the cortex and periosteal reaction There is little or no destruction of the adjacent bone The important features comprise marked density of the bony mass sharply defined borders and occasionally discrete secondary masses In differential diagnosis it is necessary to consider localized myositis ossificans, intraligamentous osteoma osteoma of the tendon sheath, and sclerosing osteogenic sarcoma

Therapy Operative removal of the entire mass is usually unsatisfactory Early amputation is indicated in most instances, although this may not be necessary if biopsy shows a definitely benign character to the tumor Amputation should be performed only on finding fibrospindle cell sarcoma in the excised tumor or after roentgen manifestations of recurrence

ADDITIONAL READING

GESCHICKTER C F and COPELAND M M Parosteal Osteoma of Bone a New Entity Ann Surg *133* 790 1951

Osteoid Osteoma

Osteoid osteoma was first described by Jaffe in 1935 It is a small round or oval nidus like lesion of bone which seldom exceeds one centimeter in diameter The lesion may lie within any portion of the affected bone Although the nidus like focus is generally small, the abnormal zone of reaction may be very large the thickening and sclerosis extending for a distance of several centimeters around the nidus The lower limbs are affected more commonly than the upper The femur is one of the most common sites The lesion may occur in the astragalus tibia fingers toes vertebral column and pelvic bones It is not definitely agreed whether it is a benign tumor of bone or represents an atypical infectious process By many it is considered to be infectious in origin, a form of sclerosing osteitis The clinical and roentgen manifestations support Jaffe's contention that it is a neoplasm The adherents of the infection theory base their conclusion on the fact that the lesion is a subcortical bone abscess which has produced minimal destruction associated with pronounced sclerosis of the adjacent bone

The course is frequently benign The history is of long duration the patient usually complaining of pain in the affected bone and local tender-

ness for months or years The pain is not severe although it may be constant and increase until it keeps the patient awake at night Swelling may be absent Lesions in the lower extremities are associated with disability and a limp Fever and erythema are unusual A history of trauma is not common The condition occurs mainly between the ages of five and twenty four years the extremes being two and fifty years About 40 per cent of the cases have been in children less than sixteen years old

Roentgen Manifestations The manifestations vary with the duration of the lesion and the bone involved In the early stages, a small area of increased density in the bone is the only finding During the intermediate stage there is a circular radiolucent area which may contain a central



Fig 295 Fig 296

Fig 295 Osteoid Osteoma

Fig 296 Osteoid Osteoma There is a localized area of sclerosis involving the lateral aspect of the upper third of the tibia (arrow) Histopathologic study confirmed the roentgen diagnosis of osteoid osteoma

lesion The fully developed process resembles an osteoma It may be necessary to take numerous oblique views or laminograms in order to demonstrate the central nidus the small dense shadow which represents ossification of the central portion of the lesion About the nidus there is a thick dense zone of sclerotic bone in practically every instance Lesions at or near the cortex are associated with extensive regional hypertrophy particularly along the periosteal surface In the presence of marked increase in the circumference of the shaft of the bone with extensive sclerosis, demonstration of the nidus becomes difficult The roentgen manifestations are characteristic and consist of a dense sclerotic, markedly radiopaque zone in the bone within which there is a small oval or rounded area of radiolucency The area of radiolucency may contain a small rounded sequestrum The outer area in some instances measures 4 cm

or more in length The nidus may appear immediately beneath the periosteum, deeper in the cortex of the bone, or within the medullary canal In the early stages, diagnosis is difficult or impossible as the reactive elements in the bone are not demonstrable by roentgen methods After the disease has been present for some time, the surrounding sclerosis may become so dense that the nidus is not demonstrable

Pathogenesis The nature, cause, and course of the condition are unknown It has been postulated that the lesion is a healing area of bone infarction The disease may undergo spontaneous healing In view of the fact that in practically all cases the lesion is discovered in patients under thirty years of age it appears that the disease has undergone spontaneous healing in many instances The picture and clinical course of the disease deviate in many respects from those generally understood to be the case with benign tumor Since the lesion may undergo spontaneous healing, it does not appear that it is caused by bone infarction The consensus is that osteoid osteoma is a benign tumor of bone and is usually of endochondral origin

Pathology Histopathologic examination shows a circular lesion 2 cm or less in diameter which is reddish brown and well demarcated from the surrounding reactive bone It is composed of osteoid tissue, condensed near the center to resemble true bone There are no evidences of inflammatory reaction Jaffe's histopathologic description of the lesion in 1935 is classical There is dense sclerotic overgrowth of bone The lesion is typically round or oval with considerable reaction about it There is a nidus which is composed of a background of vascular fibrous stroma with lymphocytic infiltration multinucleated giant cells and osteoid tissue In the early stages, the tumor is made up of a vascular mesenchymal substratum of osteoblasts This calcifies slowly and the tumor contains large amounts of osteoid or multiple osteoid trabeculae The osteoid eventually is converted into compact bone When the osteoid osteoma develops in spongy bone it is surrounded by vascular tissue and an area of sclerotic bone In compact bone the perifocal reaction is more marked The cortex is thickened over a large area and is composed of two layers Rudimentary islets of bone may be present

Differential diagnosis must include chronic osteomyelitis, the sclerosing osteomyelitis of Garre, tuberculosis atypical osteosarcoma and Ewing s endothelioma of bone The lesion is differentiated from malignant neoplasm of the bone by the well localized topographic pattern and the absence of bone destruction Treatment consists of surgical removal by block dissection if possible If this cannot be done thorough curettement is curative Roentgen therapy has been reported as effecting a cure in some cases Complete and immediate disappearance of pain occurs after surgical removal

Chondroma

Chondromas are tumors which consist primarily of cartilage The cartilage in most cases is hyaline although in some instances it is partly or entirely of the fibrous or elastic type Cartilaginous neoplasms lack one important characteristic of tumor that is unlimited growth potential They follow the general growth curve of the individual and usually cease to grow after puberty All of the bones with the exception of those of the vault of the cranium and upper portion of the face are preformed in carti

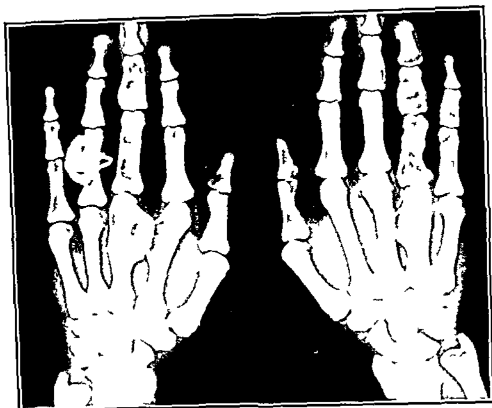


Fig 297 Multiple Enchondromas There are multiple areas of increased radiance with expansion and thinning of the cortex involving many of the bones of the hands Biopsy revealed multiple enchondromata



Fig 298 Chondroma There is an ovoid area of increased radiance involving the upper third of the diaphysis of the humerus The cortex is absent in the affected area The medial borders of the involved segment are eburnated and appear slightly lobulated The epiphysis and the epiphyseal line are not affected Histopathologic study of a specimen removed for biopsy revealed a chondroma

lage and small cartilaginous rests may persist throughout life Proliferation of the islands of cartilage results in the formation of a chondroma These neoplasms may become osteochondromas and ultimately osteomas by replacement of the cartilage by bone They may be located centrally or at the periphery of the bone When central, they are termed enchondromas The peripheral type are called ecchondromas or exostotic chondromas The cartilage of a chondroma may calcify or be replaced by bone as occurs with normal cartilage Ossification follows the same sequence as in normal endochondral ossification The pathologic entity known as hereditary exostosis or hereditary deforming chondrodysplasia must not be included in the category of chondromas In these lesions



Fig 299 Chondroma of the Ilium A Roentgenogram of the pelvis There is marked expansion of the right ilium Multiple linear striations of bone are present in the peripheral portions of the neoplasm B Two and one half years later After roentgen therapy the neoplasm has decreased in size and shows extensive recalcification

there is generalized disturbance of development of the skeleton and the cartilaginous exostosis plays merely an incidental role (see p 84)

The most frequent sites of chondromas are the bones of the fingers and toes the sternum the pelvic bones and the ribs Less commonly, chondromas occur in the vertebrae and in the metaphyseal regions of the long bones particularly the lower extremities Lesions in other bones such as the mandible are rare A chondroma frequently consists of several lobes of cartilage separated from each other by septa of connective tissue which are continuous with the capsule of the tumor The growth of the tumor is by apposition as a rule Some authorities are of the opinion that chondromas originate in remnants of the epiphyseal cartilage persisting in the



Fig 300 Chondroma of the Humerus *A* There is marked expansion and rarefaction of the upper end of the humerus The cortex is thinned being barely visible along the medial aspect of the lesion Multiple strand like areas of bony density traverse the lesion in an irregular pattern *B* Seventeen months later The lesion is more extensive than previously

spongy trabeculae of the shaft after the growth of the bone has been completed Spicules of cartilage surrounded by bone may persist deep in the shaft under normal conditions These remnants of cartilage consist of calcified intercellular substance and are not capable of proliferation Multilocular chondromas are frequent and indicate that development of new islands of cartilage from connective tissue is relatively of common occurrence They are *not derived* from supernumerary joint cartilage or persisting embryonic tissue of the precartilaginous type They may show regressive changes with calcification liquefaction and mucous degeneration which often is associated with the formation of cysts

Roentgen Manifestations The roentgen manifestations vary with the type of chondroma the enchondromas producing changes which differ from those in the peripheral or exostotic type The centrally placed chondroma or enchondroma characteristically causes marked expansion of the bone with an area of increased radiance in the bone The cortex is thinned and forms a thin border at the periphery of the lesion Multiple

bony septa traverse the affected area in the bone and small flecks of calcium occupy the central portion of the lesion. The osseous defect presents a sharply defined border which may be smooth or irregular in outline but shows no evidence of sclerosis. In the phalanges and metacarpals enchondromas may be numerous and produce an extensive area of increased radiance traversed by many fine bony septa. The bone is expanded and the cortex is thinned. The appearance is that of a cystic, multilocular process. The cortex may be expanded so markedly that it is barely visible as a thin, linear area of calcific density or disappears completely.

The chondromas which affect the cortical portion of the bone primarily are characterized by thinning and eccentric displacement of the affected portion of the cortex. There is increased radiance with almost complete absence of bone at the site of the lesion. The margins of the involved area reveal little or no evidences of sclerosis. The exostotic form is characterized by the presence of a pedunculated mass extending from the external surface of the bone. The cortex is expanded at the base of the pedicle of the neoplasm. The peripheral portion of the tumor forms a widened irregular area of radiant and mottled density within which are interspersed curvilinear and lobulated zones of calcification. This is frequently referred to as a large cauliflower-like cartilaginous and bony mass. Chondromas of the scapula, ilium, ribs, sternum and other flat bones may form large, expansile neoplasms. There is extensive rarefaction at the site of the lesion. Linear and mottled calcification is distributed irregularly within the area of rarefaction and may extend into the adjacent tissues beyond the normal confines of the bone. In the ribs and jaws fusiform expansion and localized areas of radiance are common. Pathologic fractures are frequent and are due to the decalcification, thinning and expansion of the bone. Rapid increase in the size of the lesion as shown on successive roentgen studies made at intervals of a few weeks is an important indication of malignant degeneration.

Periosteal Chondroma

Lichtenstein and Hall describe a distinctive type of tumor which they term periosteal chondroma. The periosteal chondroma represents a benign tumor of cartilage which apparently originates within and beneath the periosteal connective tissue. It has not generally been recognized in the past, although it is not rare. It grows slowly and attains only comparatively small size. The tumor characteristically erodes and induces sclerosis of the contiguous cortical bone. In the six cases described by them the lesion occurred in male and female patients and in young children and adults, sex and age apparently being of little significance. The neoplasm involved the bones of the foot and hand including the carpus and the cortex of the large limb bones, two cases having been in the finger phalanges, one in the carpal navicular bone, one in a metatarsal bone and one each in apposition to the upper shaft of the tibia and the humerus.

Clinical Manifestations. The lesion manifests itself by pain, gradual swelling and local tenderness. The duration of symptoms may range from a few months to ten years. There is generally a small slightly tender tumor which may produce limitation of motion if located near a joint. At operation there is a rubbery firm lobulated cartilaginous tumor adhering to the periosteum which is partially nestled within the gouged out under

lying cortex. The tumor tends to develop slowly and insidiously and may require a number of years to attain appreciable size. In some instances it has been present for as long as ten years. Trauma does not appear to be a significant etiologic factor.

Roentgen Manifestations. There is a comparatively small, trough-like declivity in the cortex at the site of the lesion. The base of this lunate hollow area consists of sclerotized cortical bone, apparently a reflection of the reaction to slow pressure erosion. The tumor itself produces a vaguely defined, soft tissue shadow. Within the tumor, there may be small focal areas of calcific density. In the oblique and lateral projections the

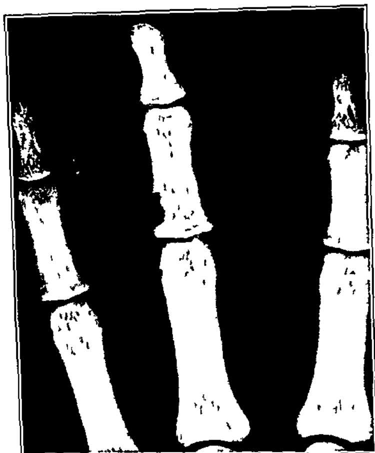


Fig. 301. Periosteal Chondroma. There is an area of destruction involving the proximal end of the middle phalanx of the middle finger. The margins of this area are lobulated, sharply defined and show slight eburnation. The roentgen diagnosis of periosteal chondroma was confirmed by histopathologic study of the lesion removed at operation.

depressed sclerotized cortical base may be superimposed on that of the shaft of the affected bone and give a false impression of a central tumor of rounded or ovoid contour such as an enchondroma. In most cases the peripheral contour of the tumor is not visualized although in rare instances it may become outlined by a delicate convex shell of periosteal bone. The diagnosis is aided by the fact that one may observe a regular focal area of increased density within the lesion nestling in the hollow of the eroded cortex, which indicates that it is probably of cartilaginous nature. In the case of involvement of the fingers one must consider in differential diagnosis the possibility of a glomus tumor or pigmented tenosynovitis, the so-called giant cell tumor of the tendon sheath, with erosion of the

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the distal epiphysis of the femur and the proximal epiphyses of the tibia and humerus Cases have been reported in the talus Pain and swelling are the most common complaints The pain is usually moderate Enlarged lymph glands may be present Pathological fractures are rare There are no general manifestations The usual laboratory tests are not helpful in diagnosis

Roentgen Manifestations The characteristic lesion comprises a round or oval, well defined area of destruction in the cancellous bone Reaction to the destruction is frequently evident Usually, the border of the defect is irregular and poorly defined The lesion is mottled in character and is described as resembling cotton The characteristic trabecular appearance and cortical expansion of giant cell tumors are absent The position of the lesion in relation to the epiphyseal plate is variable In many cases, the tumor is predominantly on the epiphyseal side It may invade both the joint and the metaphysis In other instances the tumor is situated entirely on the metaphyseal face of the epiphyseal cartilage In the case of the talus the lesion is in the body of the bone as a rule but may involve the borders of the bone Subperiosteal bone production is rare, but may occur Some observers have found that the periosteal reaction usually extends from the metaphysis to the diaphysis, providing a characteristic manifestation which distinguishes the lesion from Ewing's tumor Serial roentgenograms may reveal extension toward the joint Transarticular spread may occur with involvement of the adjacent bone

Pathology The basic cell of the tumor is a reticulohistocytic cell with a tendency to differentiation into chondroblastic tissue and cartilage The basic stromal cell rather than the giant cell establishes the difference between this group of tumors and giant cell tumors In the latter, there is fibroblastic differentiation but never sequential cartilage formation The tumor cells are described as newly formed cartilage cells associated with absorption of misplaced islands of cartilage The interpretation of the basic tumor cell as reticulohistocytic has been based primarily on the observation of the various stages of transition between the tumor cell the chondroblast, and cartilage and by the presence as demonstrated by silver stains of reticuloendothelial elements and reticulum fibers

Treatment and Prognosis The tumor appears to be benign as manifested by its course following treatment The possibility of malignant degeneration must be borne in mind Many fatal cases have been described Those which are malignant appear to be primarily metaphyseal in origin in contradistinction to the benign cases which are primarily epiphyseal Both benign and malignant varieties exist and differentiation of the two types is exceedingly difficult Conservative therapy has been curative in many cases If the lesion penetrates the epiphysis and destroys the joint cartilage it should be excised bone grafts being inserted to make up for loss of substance and to maintain the continuity of the limb When surgical approach is impossible, roentgen therapy should be utilized

Osteochondroma

Osteochondromas tend to develop at the metaphyses of the long bones or in the flat and short bones and are the result of displacement of epiphyseal cartilage to the surface of the bone The solitary lesions vary widely in size and shape They frequently occur in the region of the epiphyseal

contiguous cortex The roentgen manifestations are characteristic and establish the diagnosis with definiteness

Pathologic Findings On gross examination, the tumor is circumscribed and lobulated It may be flattened, rounded or ovoid The size varies from 1 5 to 3 5 cm in diameter Most of the lesions tend to be small On microscopic examination, the tumor is composed of lobules of hyaline cartilage with a basophilic matrix in greater or lesser part There are cartilage cell nests similar to those in enchondroma In some instances the tumor looks like a chondrosarcoma However, it is a benign tumor in the opinion of Lichtenstein and Hall

Treatment and Prognosis The best therapy appears to be conservative surgical extirpation The tumor is usually circumscribed and can be removed from its bed, although it may adhere to the periosteum Roentgen therapy does not appear indicated Surgery utilizing extirpation and curreting of the eroded sclerotized cortical bone has given satisfactory results Block excision may be feasible

ADDITIONAL READING

LICHTENSTEIN L and HALL J I Periosteal Chondroma Jour Bone & Jt Surg *34A* 691 1952

Epiphyseal Chondroblastoma of Bone

Epiphyseal chondroblastoma of bone is a neoplasm of the chondroblastic series which only recently has been recognized as an entity The clinical and histopathological characteristics of the tumor are clearly defined Correct classification is of great significance to establish the diagnosis and permit of adequate treatment In the older literature the tumor has been described under varying names Codman and Ewing termed it a chondromatous or calcifying variant of giant cell tumor Geschickter and Copeland first considered it a chondroblastic sarcoma and later termed it a benign or malignant chondroblastoma The tumor was classified as benign chondroblastoma by Jaffe and Lichtenstein It was first recognized as a distinct entity by Ewing in 1928, who termed it a calcifying giant cell tumor Codman described an epiphyseal chondromatous giant cell tumor of the upper end of the humerus and found nine cases in patients aged twelve to twenty-five in the Registry of Bone Tumors He stressed the fact that it was benign in character and presented many difficulties in differential diagnosis It is possible that many cases reported as cures of chondrosarcoma by conservative therapy are, in reality lesions of this group Jaffe and Lichtenstein were the first to suggest that these tumors were chondroblastic in origin rather than sarcomas or variants of giant cell tumors They emphasized the fact that they originate in the epiphysis of the long bones but do not show a predilection for the proximal end of the humerus as stated by other authors They also stress the fact that the treatment of choice is curettement with or without x-ray therapy

Clinical Characteristics The tumor originates during the adolescent years or just after the closure of the epiphyses has been completed Since they have their origin in the epiphyseal cartilage they do not occur after growth has ceased and the epiphyseal cartilage has disappeared The patients *range in age from thirteen to eighteen years The tumor is in* close relation to the epiphyseal line The most frequent sites have been

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Epiphyseal chondroblastoma of bone is a neoplasm of the chondroblastic series which only recently has been recognized as an entity The clinical and histopathological characteristics of the tumor are clearly defined Correct classification is of great significance to establish the diagnosis and permit of adequate treatment In the older literature the tumor has been described under varying names Codman and Ewing termed it a chondromatous or calcifying variant of giant cell tumor Geschickter and Copeland first considered it a chondroblastic sarcoma and later termed it a benign or malignant chondroblastoma The tumor was classified as benign chondroblastoma by Jaffe and Lichtenstein It was first recognized as a distinct entity by Ewing in 1928, who termed it a calcifying giant cell tumor Codman described an epiphyseal chondromatous giant cell tumor of the upper end of the humerus and found nine cases in patients aged twelve to twenty five in the Registry of Bone Tumors He stressed the fact that it was benign in character and presented many difficulties in differential diagnosis It is possible that many cases reported as cures of chondrosarcoma by conservative therapy are, in reality, lesions of this group Jaffe and Lichtenstein were the first to suggest that these tumors were chondroblastic in origin rather than sarcomas or variants of giant cell tumors They emphasized the fact that they originate in the epiphysis of the long bones but do not show a predilection for the proximal end of the humerus as stated by other authors They also stress the fact that the treatment of choice is curettement with or without x ray therapy

Clinical Characteristics The tumor originates during the adolescent years or just after the closure of the epiphyses has been completed Since they have their origin in the epiphyseal cartilage, they do not occur after growth has ceased and the epiphyseal cartilage has disappeared The patients range in age from thirteen to eighteen years The tumor is in close relation to the epiphyseal line The most frequent sites have been

Osteochondroma of the Skull. Osteochondroma of the skull is a rare condition. The most frequent location is at the base arising from the sphenoid, ethmoid and occipital bones, the bones which develop from cartilaginous matrices. The tumors may attain very large size. They usually manifest themselves clinically when the patient is about the age of twenty to thirty years. The rate of growth is slow and they may be present for as long as ten or even twenty years. If malignant degeneration occurs, rapid growth ensues. When located in the parasellar region, there is usually an early paresis of one or more of the third, fourth, fifth, and sixth nerves. Headache is common. Increased intracranial pressure is a late development because of the slowness of growth. Exophthalmos, local edema, and venous stasis of the eyelid may result from impairment of the venous return to the cavernous sinus. Visual defects with optic pallor develop if the optic nerves or chiasm are involved. A pituitary hypothalamic syndrome is seen in association with osteochondromas which encroach upon the hypophysis and hypothalamus. The temporal lobe, the cerebral peduncles and the cranial nerves numbered seven to twelve may be involved as the growth enlarges. The characteristic roentgen finding is a dense bony neoplasm with irregular margins. There is mottling within the tumor, the rarefaction being due to the fact that cartilage casts no shadow on the roentgenogram. In the basal type, there is unilateral bone destruction of the base of the skull which may involve the parasellar region, the lesser wing of the sphenoid, the petrous apex, the floor of the anterior fossa and the medial wall of the orbit.

In differential diagnosis, aneurysm of the internal carotid artery, meningioma, craniopharyngioma, chordoma, acoustic neuroma, intracranial dermoids, metastatic carcinoma, and lymphepithelioma (Schmincke's tumor) must be considered. Aneurysm is apt to present curvilinear, parallel, calcific deposits or a smooth, rounded area of calcification. Meningioma may be difficult to differentiate, and this also applies to dermoids and other neoplasms which cause bone destruction in the base of the skull and tend to calcify. The craniopharyngiomas are more apt to balloon the sella; they seldom destroy other normal bony structures, and their granular or linear calcific deposits are limited to the suprasellar or intrasellar regions. In metastatic carcinoma, Schmincke's tumor, and acoustic neuroma, there are erosions of the base and petrous apex, usually without calcific deposits.

Non-osteogenic Fibroma of Bone

Non-osteogenic fibroma of bone was first described by Jaffe. He considered it neoplastic in origin and a definite clinical entity. Hatcher, however, was of the opinion that the condition constitutes a disturbance of endochondral bone formation and represents a localized manifestation of fibrous dysplasia. The lesion is composed of spindle shaped connective tissue with multinuclear giant cells and foam cells. There is no evidence of bone formation. The process comprises an accumulation of connective tissue which has not undergone osseous metaplasia. It is a disease of young persons and is frequently asymptomatic, being discovered during the course of a roentgen examination. There may be slight pain, tenderness and local swelling in the region of the lesion. The cause of the condition is not known.

line and are usually directed away from the nearest joint The point of attachment may consist of a broad base or a long narrow pedicle The cortex of the bone is continuous along the margins of the growth, although it is thinner or entirely absent at the distal portion of the tumor The roentgen appearance is that of an osseous tumor with a dense bony base and a cauliflower like, irregularly mottled distal portion When large amounts of cartilage are present there are multiple areas of radiance scattered throughout the growth Multiple cartilaginous exostoses or multiple osteochondromas are hereditary anomalies of the skeleton There is retardation of growth of the affected bones, expansion of the metaphyses and multiple bony and cartilaginous exostoses near the ends of the shafts of the long bones The short and flat bones may also present exostoses The lesions never occur in bones which are preformed in membrane or at the epiphyses *The disease begins in early childhood, progresses until*



FIG 302 Osteochondroma *A* Anteroposterior view *B* Lateral view There is a large neoplasm in the infratrochanteric region The lesion presents the characteristic manifestations of an osteochondroma

adolescence and remains unchanged after full growth has been attained Interference with growth of the long bones is usually irregular The ulna is short and there is curvature of the radius with ulnar deviation of the wrist and hand the so called Madelung s deformity The fibula may be foreshortened with lateral displacement and angulation of the astragalus In the presence of exostoses the ends of the shafts of the involved bones are widened particularly the upper humerus lower radius and similar portions of the bones at which the longitudinal growth is greatest The exostoses vary in size from a few millimeters to many centimeters in length and form as spurs with bone formation similar to the parent bone from which the neoplasm has originated The spurs point away from the nearest joint Sarcomatous degeneration may occur In adult life, the *cartilage at the distal portion of the exostoses diminishes Fracture of* the pedicle may be associated with non union and apparent decrease in the size of the exostosis The process probably represents a growth disturbance rather than a true neoplasm formation

On roentgen examination there is an ovoid or rounded area of increased radiance in the bone usually near the extremity of a long bone The lower extremity is more commonly involved While usually solitary multiple lesions may occur The margins of the defect are loculated and may be increased in density Within the lesion there are multiple thin septa or complete absence of bone structure As a rule the process is in the peripheral portion of the bone It may involve the entire width of the bone and be associated with slight bulging The cortex is thinned expanded, or completely destroyed There is no periosteal reaction or other change in the adjacent soft tissues In differential diagnosis it is necessary to consider bone cyst hemangioma sarcoma osteomyelitis and similar lytic bone diseases Therapy consists of local excision Cure is complete and there is no tendency to recurrence after surgical removal of the affected area

Giant Cell Tumor of Bone

Giant cell tumor of bone is usually classified as a benign neoplastic process which may occur in practically any bone of the body It is also known by the name osteoclastoma or benign myeloid sarcoma Typically the giant cell tumor involves the epiphysis of a long bone In the early stages it may be found in the metaphysis and prior to closure of the epiphyseal plate the growth is in some instances confined to the meta physis After the epiphyseal line has closed, there is rapid extension into the epiphysis to the articular cartilage The joint is not crossed as a rule The roentgen manifestations are characteristic The bone is widened the periosteal outline is preserved the lesion is sharply demarcated at the diaphyseal border, there is a multicystic appearance and there is no evidence of periosteal elevation or proliferation The process involves most of the width of a cancellous bone The margins are sharp and may be scalloped Numerous fine bony trabeculæ traverse the involved area The cortex is thin and bulges outward Advancement of the growth is by the lines of least resistance along the shaft and to the joint surface Extension into the femoral condyles, transverse processes of the vertebræ and other bony projections is common The lesion begins centrally and extends more to one side than the other eventually affecting the entire width of the bone While the trabeculated appearance is commonly seen lytic lesions with absorption of large areas of bone also occur Progression is manifested by increasing osteolysis with increase in the size of the bone defect and destruction of the cortex The tumor is usually solitary Pathologic fractures may occur Spontaneous healing or malignant degeneration may ensue

The roentgen manifestations are entirely destructive in character the destruction being stimulated by neoplastic osteoclasts in the portion of the bone which is under normal conditions the center of osteoclastic activity Slow growing lesions have a dense sclerotic margin while those which are growing rapidly present a moth eaten, irregular border similar to that in sarcoma Expansion of the bone associated with a well established giant cell tumor produces the classic loculated appearance This is due to the presence of coarse bands of newly formed enveloping cortex visualized through the tumor There are no actual trabeculæ within the tumor although the roentgen appearances are those of multiple trabeculations In the case of the small bones the lesion may be expansile or confined and destructive the changes closely simulating those in enchondroma Benign giant cell tumors may occur in the vertebræ the

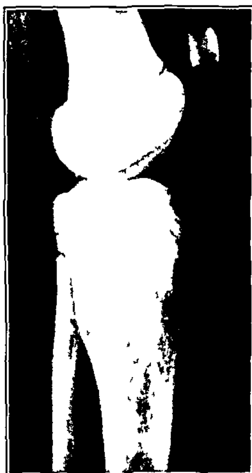


Fig 303 Non osteogenic Fibroma There is a multilocular area of increased radiance in the upper third of the diaphysis of the tibia The margins of the zone of rarefaction are sharply defined and slightly eburnated The cortex of the posterior aspect of the tibia at the site of the lesion is expanded and thinned The lesion was asymptomatic Its presence was discovered after roentgen examination of the knee subsequent to a traumatism The roentgen diagnosis of non osteogenic fibroma was confirmed by histopathologic study of a specimen removed for biopsy



Fig 304 Non-osteogenic Fibroma of Tibia There is a multilocular area of radiance in the anteromedial aspect of the lower third of the shaft of the tibia The radiant area has sharply defined, eburnated margins Histopathologic study of a specimen removed for biopsy confirmed the roentgen diagnosis of non osteogenic fibroma

On roentgen examination, there is an ovoid or rounded area of increased radiance in the bone, usually near the extremity of a long bone. The lower extremity is more commonly involved. While usually solitary multiple lesions may occur. The margins of the defect are loculated and may be increased in density. Within the lesion there are multiple thin septa or complete absence of bone structure. As a rule, the process is in the peripheral portion of the bone. It may involve the entire width of the bone and be associated with slight bulging. The cortex is thinned, expanded, or completely destroyed. There is no periosteal reaction or other change in the adjacent soft tissues. In differential diagnosis it is necessary to consider bone cyst, hemangioma, sarcoma, osteomyelitis and similar lytic bone diseases. Therapy consists of local excision. Cure is complete and there is no tendency to recurrence after surgical removal of the affected area.

Giant Cell Tumor of Bone

Giant cell tumor of bone is usually classified as a benign neoplastic process which may occur in practically any bone of the body. It is also known by the name osteoclastoma or benign myeloid sarcoma. Typically, the giant cell tumor involves the epiphysis of a long bone. In the early stages, it may be found in the metaphysis and prior to closure of the epiphyseal plate the growth is in some instances confined to the metaphysis. After the epiphyseal line has closed, there is rapid extension into the epiphysis to the articular cartilage. The joint is not crossed as a rule. The roentgen manifestations are characteristic. The bone is widened, the periosteal outline is preserved, the lesion is sharply demarcated at the diaphyseal border, there is a multicystic appearance, and there is no evidence of periosteal elevation or proliferation. The process involves most of the width of a cancellous bone. The margins are sharp and may be scalloped. Numerous fine bony trabeculae traverse the involved area. The cortex is thin and bulges outward. Advancement of the growth is by the lines of least resistance along the shaft and to the joint surface. Extension into the femoral condyles, transverse processes of the vertebrae and other bony projections is common. The lesion begins centrally and extends more to one side than the other, eventually affecting the entire width of the bone. While the trabeculated appearance is commonly seen, lytic lesions with absorption of large areas of bone also occur. Progression is manifested by increasing osteolysis with increase in the size of the bone defect and destruction of the cortex. The tumor is usually solitary. Pathologic fractures may occur. Spontaneous healing or malignant degeneration may ensue.

The roentgen manifestations are entirely destructive in character, the destruction being stimulated by neoplastic osteoclasts in the portion of the bone which is under normal conditions the center of osteoclastic activity. Slow growing lesions have a dense, sclerotic margin, while those which are growing rapidly present a moth-eaten irregular border similar to that in sarcoma. Expansion of the bone associated with a well established giant cell tumor produces the classic loculated appearance. This is due to the presence of coarse bands of newly formed enveloping cortex visualized through the tumor. There are no actual trabeculae within the tumor, although the roentgen appearances are those of multiple trabeculations. In the case of the small bones the lesion may be expansile or confined and destructive, the changes closely simulating those in enchondroma. Benign giant cell tumors may occur in the vertebrae, the



Fig 305 Giant Cell Tumor A Anteroposterior view B Lateral view The lesion was discovered during roentgen examination after a trauma The patient a twenty one year old female had no symptoms and was unaware of the presence of the lesion Histopathologic study of the specimen removed at operation confirmed the roentgen diagnosis of benign giant cell tumor

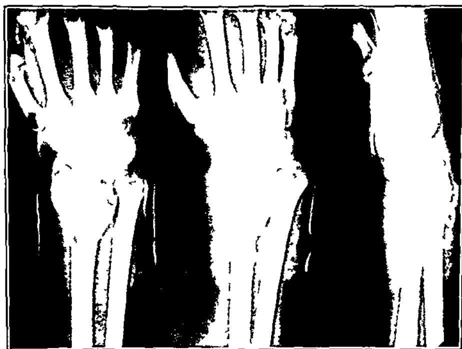


Fig 306 Giant Cell Tumor There is an area of increased radiance in the lower end of the radius The cortex of the bone is markedly expanded and thinned Multiple fine bony septa are visible within the lesion The process extends to the articular surface of the lower end of the radius There is marked osteoporosis of the bones of the wrist and hand Biopsy report benign giant cell tumor

skull, and other bones. Tumors in the long bones as well as those in the short bones may in some instances present enchondroma elements in addition to giant cell formations. These lesions are usually in the metaphysis and are associated with clubbing and widening of the metaphysis. Although considered a benign lesion the neoplasm may become malignant and metastasize, particularly to the lung. The benign and malignant forms produce identical roentgen changes and cannot be differentiated from each other. The characteristic multilocular appearance, localized destruction, irregular margins and, in some instances, a slightly sclerotic border are present in both varieties of the disease. Serial studies continued during a period of months or years may be necessary to demonstrate the presence of malignant changes.

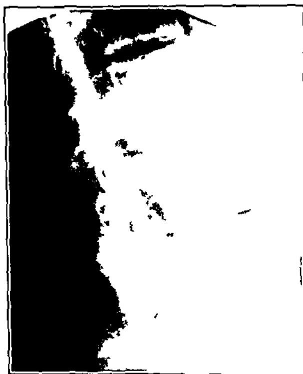


FIG 307. Giant Cell Tumor of Third Lumbar Vertebra. The third lumbar vertebra shows extensive rarefaction. The rarefied area is traversed by multiple thin irregular trabeculations. The changes involve the body, pedicles, laminas and spinous process. *Histopathologic examination of a specimen removed for biopsy showed giant cell tumor.*

There is lack of general agreement among pathologists as to what actually constitutes giant cell tumor of bone. It is important to separate true giant cell from the many variants which resemble it but are definitely different. Some of the entities which are *confused with* giant cell tumor are non osteogenic fibroma of bone, chondroblastoma of bone, aneurysmal bone cyst and the so called giant cell *tumors* of synovial or tenosynovial tissues which are known under the terms of pigmented villonodular synovitis, bursitis and tenosynovitis. *Recently* many advances have been made in our knowledge of giant cell tumor of bone. Progress has been made in establishing the distinction between the lesions of hyperparathyroidism and giant cell *tumors*. A wide variety of unrelated and generally less serious skeletal lesions have been arbitrarily associated with giant cell tumor in the guise of variants. In the past it has not been appreciated that certain giant cell tumors have a very serious prognosis.

Giant cell tumor of bone is considered a distinctive neoplasm arising from the nonbone forming tissue of the marrow The tumor is composed of a vascularized network of spindle shaped or ovoid stromal cells, heavily interspersed with multinuclear cells, apparently syncytial stromal cells The true giant cell tumor must be differentiated from its spurious variants with which it is frequently confused The so called variants have little in common clinically and anatomically with each other except as regards prognosis The variants of giant cell tumor are distinct clinical and pathological entities The so called spindle cell, healing, or xanthic variant is a non osteogenic fibroma of bone and the calcifying or chondromatous giant cell tumor is a benign chondroblastoma The latter is a benign tumor derived from cartilage forming connective tissue and is not related histogenetically to giant cell tumor There is also a cartilaginous variant of the genuine osteoclastoma Another lesion which has been mistaken for a variant of the giant cell tumor is the aneurysmal bone cyst The so called giant cell tumors of synovial or tenosynovial tissues are entirely unrelated to giant cell tumors of bone These tumors should be classified under the heading of pigmented villonodular bursitis and tenosynovitis

The genuine giant cell tumor, in contrast to many of the so called variant lesions is seldom observed in patients below the age of twenty When present in a long bone it usually involves the end of the bone Lesions which develop in the metaphyses in the great majority of cases prove to be nonosteogenic fibroma, bone cyst chondromyxoid fibroma and other less serious lesions The aneurysmal bone cyst is particularly apt to develop in the bones of the upper limbs, the skull the pelvis and the vertebrae In the past it has been stated that irradiation is effective in the treatment of giant cell tumors However many of the cases termed giant cell tumors represented epulides while others were in children and adolescents or were located in the shafts of the long bones Central nontrabeculated cystic tumors in the classical sites at the upper end of the tibia and the lower end of the femur which respond poorly to radiation are usually not giant cell tumors

Treatment The true giant cell tumor is a formidable neoplasm While not necessarily sarcomas, they are not benign in every instance Many giant cell tumors may be treated successfully by curettement or irradiation some are resistant and tend to recur Certain of these tumors behave like frank sarcomas particularly after one or more local recurrences It is believed that half of the giant cell tumors have a favorable outcome if properly treated approximately one third are resistant and recur after treatment and approximately 15 per cent are frankly malignant and *prone to metastasize to the lungs An occasional giant cell tumor is found* to be malignant at the time of the first examination More commonly, it appears that malignant degeneration ensues incidentally to one or more local recurrences It is advisable to divide the tumors with a view to prognosis into grades 1 2 and 3 according to insignificant or pronounced atypism of the stromal cells Grading is of practical value particularly in prognosis Roentgen interpretation alone cannot be relied upon in determining the type and prognosis of the tumor Treatment can only be predicated upon pathological study As a rule it is not advisable to combine surgery and roentgen therapy There is great disagreement as to which form of therapy is preferable Whenever it is possible to excise or resect the tumor *in toto* this is the method of choice as it is the most

certain way to effect a cure. If the tumor is large and involves the lower end of the femur or the upper end of the tibia, resection is usually not possible. In these cases curettement is the procedure of choice. However, residual tumor tissue adheres to the walls of the cavity and results in recurrence. Other forms of treatment include chemical cauterization of the walls of the cavity and packing the cavity with bone chips to hasten osteogenesis.

Solitary Unicameral Bone Cyst

Solitary unicameral bone cyst is a distinctive lesion. While relatively rare, it is frequently met in childhood and adolescence. Cysts of this type are most common in the long bones and usually are located in the proximal third of the diaphysis or shaft of the humerus, femur, and tibia.



Fig 308 Fig 309

Fig 308. Bone Cyst with Pathologic Fracture. There is a large cyst in the upper third of the diaphysis of the humerus with a comminuted fracture in the region of the cyst. The fracture resulted from a minor trauma.

Fig 309. Bone Cyst. The upper end of the diaphysis of the humerus presents a typical bone cyst.

The jaw and other flat bones may be involved. The cyst may be present for long periods of time and attain large size prior to being discovered. A slight trauma may result in a pathologic fracture of the cyst wall and in many instances the lesion is not recognized until this sequence of events happens. There may be a history of localized or radiating pain aggravated by exercise. In most cases the cysts occur in the age groups between three years and twenty years. Patients in the older age groups may also show this lesion. The conditions which may closely simulate bone cyst comprise giant cell tumors, hyperparathyroidism, polyostotic fibrous dysplasia, solitary plasmacytoma, eosinophilic granuloma, neurofibroma, primary or metastatic bone tumor, and xanthomatosis.

In many instances the roentgen manifestations are not characteristic and final diagnosis must rest on a biopsy. It is characterized by a sharply

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Treatment. The true giant cell tumor is a formidable neoplasm. While not necessarily sarcomas, they are not benign in every instance. Many giant cell tumors may be treated successfully by curettement or irradiation; some are resistant and tend to recur. Certain of these tumors behave like frank sarcomas, particularly after one or more local recurrences. It is believed that half of the giant cell tumors have a favorable outcome if properly treated, approximately one-third are resistant and recur after treatment, and approximately 15 per cent are frankly malignant and prone to metastasize to the lungs. An occasional giant cell tumor is found to be malignant at the time of the first examination. More commonly it appears that malignant degeneration ensues incidentally to one or more local recurrences. It is advisable to divide the tumors with a view to prognosis into grades 1, 2, and 3 according to insignificant or pronounced atypism of the stromal cells. Grading is of practical value, particularly in prognosis. Roentgen interpretation alone cannot be relied upon in determining the type and prognosis of the tumor. Treatment can only be predicated upon pathological study. As a rule it is not advisable to combine surgery and roentgen therapy. There is great disagreement as to which form of therapy is preferable. Whenever it is possible to excise or resect the tumor in toto, this is the method of choice, as it is the most

certain way to effect a cure. If the tumor is large and involves the lower end of the femur or the upper end of the tibia, resection is usually not possible. In these cases curettement is the procedure of choice. However residual tumor tissue adheres to the walls of the cavity and results in recurrence. Other forms of treatment include chemical cauterization of the walls of the cavity and packing the cavity with bone chips to hasten osteogenesis.

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FIG. 308 FIG. 309

FIG. 308. Bone Cyst with Pathologic Fracture. There is a large cyst in the upper third of the diaphysis of the humerus with a comminuted fracture in the region of the cyst. The fracture resulted from a minor trauma.

FIG. 309. Bone Cyst. The upper end of the diaphysis of the humerus presents a typical bone cyst.

The jaw and other flat bones may be involved. The cyst may be present for long periods of time and attain large size prior to being discovered. A slight trauma may result in a pathologic fracture of the cyst wall and in many instances the lesion is not recognized until this sequence of events happens. There may be a history of localized or radiating pain aggravated by exercise. In most cases the cysts occur in the age groups between three years and twenty years. Patients in the older age groups may also show this lesion. The conditions which may closely simulate bone cyst comprise giant cell tumors, hyperparathyroidism, polyostotic fibrous dysplasia, *solitary plasmacytoma*, eosinophilic granuloma, neurofibroma, primary or metastatic bone tumor and xanthomatosis.

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Fig. 308 Fig. 309

Fig. 308. Bone Cyst with Pathologic Fracture. There is a large cyst in the upper third of the diaphysis of the humerus with a comminuted fracture in the region of the cyst. The fracture resulted from a minor trauma.

Fig. 309. Bone Cyst. The upper end of the diaphysis of the humerus presents a typical bone cyst.

The jaw and other flat bones may be involved. The cyst may be present for long periods of time and attain large size prior to being discovered. A slight trauma may result in a pathologic fracture of the cyst wall and in many instances the lesion is not recognized until this sequence of events happens. There may be a history of localized or radiating pain aggravated by exercise. In most cases the cysts occur in the age groups between three years and twenty years. Patients in the older age groups may also show this lesion. The conditions which may closely simulate bone cyst comprise giant cell tumors, hyperparathyroidism, polyostotic fibrous dysplasia, solitary plasmacytoma, eosinophilic granuloma, neurofibroma, primary or metastatic bone tumor and xanthomatosis.

In many instances the roentgen manifestations are not characteristic and final diagnosis must rest on a biopsy. It is characterized by a sharply

defined oval or rounded area of increased radiance with its long diameter extending along the shaft The cortex is thinned and may be expanded The lesion is not trabeculated or at most shows a few fine trabeculae Spontaneous fracture is common and may be the first indication of the existence of this lesion Healing is prompt with much callus formation and usually results in disappearance of the cyst

Aneurysmal Bone Cyst

Aneurysmal bone cyst is a distinct clinical roentgen and pathologic entity first presented by Jaffe and Lichtenstein in 1942 It is an essentially solitary, localized and expanded fibrous lesion honeycombed by an enormously dilated, plexiform vascular bed The disease has been described in the literature under a variety of names such as primary giant



Fig 310 Aneurysmal Bone Cyst A Lateral projection There is an extensive area of rarefaction in the temporoparietal region The margins of the affected area are eburnated irregular in outline and scalloped There are multiple bony trabeculae in the area of rarefaction which divide the lesion into multiple loculi *B* Posteroanterior projection There is marked rarefaction and expansion of the bones in the affected area Multiple loculi are clearly demonstrable The tables are thinned and expanded

cell tumor subperiosteal giant cell tumor giant cell variants of bone cysts osteitis fibrosa cystica pulsating benign giant cell tumor angioma ossifying hematoma and osteogenic sarcoma The exact nature of the lesion has not been established and there is no agreement as to whether it is actually a neoplasm Jaffe and Lichtenstein adopted the name "aneurysmal bone cyst because of the bulging of the bone and the fact that the lesion is composed of a group of blood filled spaces of varying size The clinical manifestations are variable There may be pain swelling tenderness and limitation of motion As with other bone neoplasms there is not infrequently a history of a localized trauma preceding the onset of symptoms Cord compression may ensue in lesions of the vertebrae The disease occurs predominantly in persons under twenty years of age It appears benign Simple curettage usually effects a cure and surgery is the therapy of choice provided the cyst is in an accessible

location. Amputation is not necessary and radical surgical procedures are contraindicated. Roentgen radiation may effect a cure.

Roentgen Manifestations. The lesion is most common in the long bones and the vertebræ. The flat bones, the calvaria, the tarsal bones and the metacarpals may also be affected. The condition is characterized by an eccentrically placed defect and causes ballooning of the cortex on the affected side. There is a cyst like area of radiance with, in many instances, multiple trabeculæ. The cortex is thinned and the margin of the lesion presents a sharply defined border of periosteal bone. In the case of the long bones, the affected area is near one end of the shaft. The epiphysis is usually not involved. Cysts of the vertebrae may be in the body or neural arch and, as the neoplasm increases in size, an adjacent vertebra or rib may be implicated with resultant compression of the cord. Many cases are atypical and present alterations which differ to a considerable degree from the so called typical picture. There is expansion of the bone, often marked in degree, and cystic transformation within the involved area. In the long bones, the expanding lesion extends eccentrically, which is usually not the case in giant cell tumor, and involves the internal part of the bone for only a portion of its width. The flat bones may show involvement of the entire width of the bone with a fusiform, symmetrical expansion. The expanding lesion usually assumes an ovoid shape with the axis in the direction of the bone within which it lies. The size of the cyst formation varies widely. In the vertebral arch, the lesion may measure only 2 to 3 cm. in diameter, while in the long bones it may attain a size of 8 to 10 cm. in its long diameter. The interior of the affected area shows complete replacement of the normal architecture by a cavernous fibro vascular tissue. Multiple pools of blood are contained in dilated preformed vascular channels which are delimited by connective tissue. There is hemorrhage, giant cell reaction, and reparative new bone formation in the connective tissue. Jaffe has reported 20 cases of this type of bone cyst. Roentgen examination raises the suspicion of a malignant neoplasm, particularly in the minds of observers unfamiliar with this type of cyst.

Differential Diagnosis. It is necessary in differential diagnosis to consider malignant neoplasm of bone, enchondroma, hemangioma, myeloma, fibrous dysplasia, hydatid disease of bone, and other lytic bone lesions. Simple bone cyst, non osteogenic fibroma and giant cell tumor may also produce closely similar changes. Benign giant cell tumors seldom occur in the vertebræ, although the sacrum may be the site of this lesion, usually affect the epiphysis, occur in persons over twenty years of age, tend to recur and, in about 15 per cent of the cases, undergo malignant degeneration *and metastasize*.

Hemangioma of Bone

Hemangioma is a neoplasm composed of newly formed blood vessels. The common sites are the vertebræ, skull, and long bones. It may occur at any age. The sex distribution is approximately equal. There are two common histologic types. Capillary hemangiomas consist of small vessels and capillaries. Cavernous hemangiomas are comprised of large thin walled vessels and sinuses lined with endothelium and containing blood. There is a connective tissue stroma in both varieties. Hemangiomas are mesenchymal in origin and probably arise from embryonal

defined, oval or rounded area of increased radiance with its long diameter extending along the shaft The cortex is thinned and may be expanded *The lesion is not trabeculated or at most shows a few fine trabeculæ* Spontaneous fracture is common and may be the first indication of the existence of this lesion Healing is prompt with much callus formation and usually results in disappearance of the cyst

Aneurysmal Bone Cyst

Aneurysmal bone cyst is a distinct clinical, roentgen and pathologic entity first presented by Jaffe and Lichtenstein in 1942 It is an essentially solitary, localized and expanded fibrous lesion honeycombed by an enormously dilated plexiform vascular bed The disease has been described in the literature under a variety of names such as primary giant



FIG 310 Aneurysmal Bone Cyst *A* Lateral projection There is an extensive area of rarefaction in the temporoparietal region The margins of the affected area are eburnated irregular in outline and scalloped There are multiple bony trabeculæ in the area of rarefaction which divide the lesion into multiple loculi *B* Posteroanterior projection There is marked rarefaction and expansion of the bones in the affected area Multiple loculi are clearly demonstrable The tables are thinned and expanded

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the center of the tumor is soft and pulsating while the periphery consists of a ring of hard bone raised above the level of the neighboring bones There is no dilatation of the vessels in the soft tissues over the lesion A bruit is usually not heard There is no alteration of hair growth in the scalp overlying the neoplasm In females there may be headaches which are paroxysmal and related to the onset of menstruation The tendency is toward external rather than internal expansion and neurological signs and symptoms are unusual When the lesion involves the orbit and the petrotemporal region the local signs and symptoms may appear much earlier with diplopia and other visual disturbances Those in the petrotemporal region may cause tinnitus and facial paresis Hemangiomas in the petrous regions may encroach on the external auditory meatus and be associated with a mass which is visible on otoscopic examination If the labyrinth is involved, there may be giddiness ataxia nystagmus and other signs of vestibular disturbances

Roentgen Manifestations The roentgen picture is that of a rounded or oval area of increased radiance with sharply defined borders Eburnation of the margins may occur but is not seen as a rule The lesion is honeycomb in appearance with numerous fine spicules radiating in more or less regular fashion from the central to the peripheral portions of the involved area The spicules are not at right angles to the bone or irregular in distribution as in the meningiomas or osteogenic sarcomas The tables are thinned and expanded The periosteum is elevated but



Fig 311 Cavernous Hemangioma of the *Frontal Bone* *A* Lateral roentgenogram of the skull There is a large rounded area of radiance in the frontal bone The margins of the defect show no evidences of eburnation Within the area of radiance there is irregular granularity with a honey-comb appearance *B* Spot film with *soft tissue* technique and a very small cone focused directly over the lesion There are irregular bony spicules extending at right angles to the bone This change is pathognomonic of hemangioma *C* Roentgenogram of a fragment of bone removed at operation The spicule formation and rarefaction of the bone characteristic of cavernous hemangioma are well shown

remnants by a process of endothelial proliferation and differentiation into blood vessels. They are benign and spontaneous regression with disappearance sometimes ensues. Malignant angiomas, termed angiosarcomas, are rare and are characterized by rapidity of growth, invasion of adjacent tissues, and metastatic involvement.

Clinical Aspects. The clinical manifestations are dependent on the site of the tumor, the rapidity of growth, and the presence or absence of malignant changes. In the spine, neurological signs and symptoms may be present. These are due to enlargement or collapse of the vertebral body, peridural extensions, or pressure on the medulla and nerve roots. In many instances, there are no associated symptoms and the lesion is discovered as an incidental finding during roentgen examination.

Roentgen Features. In the long bones, there are usually cystic changes with accentuation of the trabeculae which closely simulate those in giant cell tumor and osteitis fibrosa cystica. Small, sharply defined areas of increased radiance adjacent to the periphery of the lesion indicate progression. In the flat and irregular bones, the lesion is characterized by an area of increased radiance with coarse trabeculae extending from the center of the lesion, in some cases at right angles to the plane of the bone. In many instances the final diagnosis must await biopsy. Any bone in the skeleton may be the site of a hemangioma. The proliferation of blood vessels may start in the cortex, although usually it originates in the bone marrow. The tumor may cause extensive osseous destruction, particularly in the case of the long bones. Compensatory formation of new bone results in a fusiform swelling. In some cases the tumor destroys or weakens the vertebral body so markedly that compression fracture ensues with resultant damage to the spinal cord.

Therapy. Surgery is often attended by severe bleeding. Radiation therapy is usually successful and should be utilized except in cases with compression myelitis or other complication which necessitates prompt intervention.

Because of the marked variability and peculiar characteristics of hemangiomas of the skull and spine these must be discussed separately.

Hemangioma of the Skull

Hemangiomas of the skull present specific characteristics. The diagnosis can be established with definiteness on the basis of the roentgen manifestations. These tumors comprise about 0.2 per cent of all osseous neoplasms and about 10 per cent of the primary benign tumors of the skull. The disease has been reported in every age group, the oldest patient being seventy years and the youngest three weeks. *The highest incidence is in the fourth decade.* The lesion is three times more frequent in females than in males. There is no correlation between age and sex. Hemangiomas have been described in practically all the bones of the skull but appear to have a special predilection for the parietal bones. In rare instances the condition may be multiple. Several authors have noted an apparent relationship to the centers of ossification.

Clinical Manifestations. A hard lump may be present for months or years. In most instances the growth is asymptomatic and is discovered during roentgen examination. When a mass is present it is not tender, feels hard, and the soft tissues overlying it move freely. In some instances

aneurysm. In this lesion the erosion occurs from within outward and there is no expansion of the diploe or striations.

Sinus Pericranii

Sinus pericranii is a specific type of cavernous hemangioma of the pericranium in which one or more emissary vessels connect with the large intracranial blood sinuses through abnormal foramina in the skull. When associated with hemangiomas of the cerebellum or retina it may be part of the Hippel-Lindau syndrome. It is characterized by a soft, fluctuant swelling which enlarges with increase in the blood supply to the head when the head is lowered and on coughing, crying or straining. The mass is compressible and is reduced in size or disappears completely when pressure is exerted over it or the head is elevated. A bluish color may be visible in the skin overlying the swelling. The condition usually causes few if any symptoms. A thrill and a bruit may be present. The age incidence is from birth to late maturity. The lesion is slowly and gradually progressive, in many instances first being noted as a small nodule and gradually enlarging to a mass several centimeters in diameter. The etiology is unknown.

Pathologically, the process is of the nature of a cavernous hemangioma. It is usually circumscribed and limited to the pericranium. There are small foramina piercing the skull with anomalous emissary veins extending from the blood spaces of the tumor to one of the intracranial venous sinuses. Subsequently other fistulas form, the dilatation extends to the adjacent diploic veins, and the pressure of the enlarged vessels produces local bone absorption. Therapy consists of excision of the mass and electrocoagulation of the emissary veins.

Roentgen Findings. On roentgen examination, bone changes may be entirely absent. In some instances there is an area of radiance at the site of the mass with complete or partial absence of the bone and widening of the diploe at the margins of the bone defect. There may be honeycombing or irregular destruction of the underlying bone. A tortuous, dilated vascular channel may be present in the bone in the region of the soft tissue swelling. Both tables of the skull may show localized erosion. With low-kilovoltage roentgenograms, the rays being directed tangentially through the affected area, the soft-tissue mass may be demonstrable with the patient in the recumbent position. The lesion is most often seen at the sagittal suture, the occiput, and the forehead. Phleboliths may be present in the soft tissue mass in some instances.

Differential Diagnosis. In differential diagnosis, the principal conditions to be considered are meningocele, sebaceous cyst, and other soft-tissue neoplasms of the scalp. Meningocele usually occurs in the mid line and is associated with a rounded or oval defect in the underlying bone.

Hemangioma of the Spine

Hemangioma of the spine is of extreme importance as it may cause compression of the spinal cord. In many instances it is encountered as an incidental roentgen finding. In a study of 3,829 spinal columns at necropsy, Schmorl found hemangiomas in 10.7 per cent of the cases. The incidence was much greater in females than in males. In most cases the

intact Prominence of the blood vessels in the region of the hemangioma usually does not occur although there are exceptions to this rule The absence of enlarged vascular channels is an important aid in excluding the diagnosis of meningioma The development of large arterial channels adjacent to a hemangioma should raise the possibility of malignant degeneration While the usual sagittal and lateral views may suffice lesions in the occipital and petrous bones require the use of the Towne, Stenvers or Schuller's position The presence of a honeycomb appearance is pathognomonic In rare instances the derivation from a diploic vein may be apparent from the roentgenogram In the tangential projection, an important characteristic feature may be demonstrable and consists of the so called sunray appearance common to all hemangiomas of bone and due to the radiating bone trabeculae The radial disposition of the trabeculae may sometimes be apparent on direct view of the tumor although more often there is merely an irregularly fenestrated meshwork Expansion of the calvaria is an important manifestation Erosion of the outer table alone is common the inner table being preserved Ballooning of the inner table and erosion may occur

Differential Diagnosis In the differential diagnosis many conditions must be mentioned the chief of which are meningioma, sarcoma, osteitis fibrosa, fibrous dysplasia epidermoid, enlarged pacchionian depressions leptomeningeal cyst, osteomyelitis, and the anemias Meningiomas do not present a honeycomb appearance although they may produce stippled and eroded areas The vertical striations occurring in meningioma are usually parallel with each other not radially disposed as in the hemangiomas The absence of reactive sclerosis or hyperostosis is an important distinguishing characteristic In meningiomas the erosion of the skull occurs from within outward so that the internal table is affected more than the external table The presence of enlarged vascular channels and diploic lakes in relation to meningiomas is important in differential diagnosis An epidermoid of the skull may cause confusion in diagnosis In epidermoid the margins of the defect are serpiginous and sclerotic There are no striations and no honeycomb appearance The lesion is generally found in the temporal bone and only rarely involves the bones of the vault Osteomyelitis may produce diffuse mottling with multiple osseous defects There is usually no lump on the skull except in the case of Parrot s node which occurs in syphilitic lesions of the skull There is absence of striation Xanthomatosis Schuller-Christian disease, produces rarefied areas in the vault The patches are multiple and without honeycomb appearance The edges of the defects are clear and sharply defined Osteoporosis circumscripta involves the outer table first, has no radial striations and there is no honeycomb appearance This is also true of osteitis fibrosa or fibrous dysplasia

In the anemias there may be localized areas of thickening in the skull with striations in profile In Cooley s anemia there is an increase in the diploe with thinning of the outer table The changes are usually diffuse and bilateral and there are no circumscribed areas of erosion The osteolytic type of osteogenic sarcoma may be associated with a localized pulsatile swelling In this instance the inner and outer tables of the skull are involved equally and there is marked internal as well as external expansion of the skull When striations are present they are not clear-cut and there is no honeycomb appearance Other less common causes of erosion of the skull may be intracranial hemangioma so called cirsoid

diagnostic value With compression of the spinal cord the spinal fluid may show xanthochromia and elevated protein levels A positive Queckenstedt sign can usually be demonstrated

Roentgen Manifestations In the presence of involvement of the entire vertebral body, the roentgen manifestations are typical There is a reduction of bone density between the parallel vertical trabeculae, which are increased in density A *honeycomb type of rarefaction or a series of* vertical striations are present throughout the affected vertebral body and in many instances in the pedicles and transverse process The changes are due to irregular absorption and coarsening of the trabeculae occurring simultaneously in adjacent portions of the vertebra The vertebra may be normal in size convex or compressed The characteristic roentgen manifestations are trabeculae of increased bony density contrasting with intervening areas of reduced density The trabeculae are arranged as parallel vertical columns which may be connected by narrow horizontal bands In some instances the striated or net like markings are less regular and are replaced by small vacuoles in close approximation imparting a stippled appearance While the form of the affected vertebra is usually retained the margins may be displaced outward Occasionally the vertebra may be collapsed and the upper and lower limits become concave When the vertebral arches and apophyses are involved they retain their shape or become widened, thickened and deformed In these instances the striated markings may appear as cystiform shadows The intervertebral discs are generally intact Angiomas may be present in other bones or there may be associated cutaneous hepatic or splenic angiomas Pregnancy and endocrine factors appear to be significant as there is greater frequency of the lesions in females and at puberty

Differential Diagnosis In differential diagnosis it is necessary to consider Pott s disease infectious spondylitis syphilis metastatic neoplasm myeloma lymphoblastoma, Paget s disease actinomycosis and echinococcus cyst A partially calcified lesion may simulate Paget's disease However, the latter is usually ruled out by skeletal surveys and alkaline phosphatase determination It is also necessary to consider Hodgkin s disease, myeloma, and chordoma in differential diagnosis

Therapy Surgery and roentgen therapy are the usual modes of treatment Because of the great vascularity of the tumor the surgical mortality is high in some series being 20 to 25 per cent There has been objection to the use of roentgen therapy in that it involves delay in relieving compression of the cord and further damage may result Laminectomy may be necessary for decompression in the presence of sudden spinal cord compressions secondary to the lesion If irreversible damage has developed in the cord no form of therapy is of avail and roentgen therapy will not result in recovery or improvement The roentgen picture may change little if at all after irradiation although in some instances calcification of the lesion occurs

Tumors of the Mandible

The neoplasms of the jaw are best considered as a separate group for the reason that they present certain special characteristics not usually found in other regions of the body

hemangiomas had given no hint of their presence prior to the autopsy examination He noted that they tended to develop with increasing age or if present at birth increased in size later in life The most frequent sites are between the twelfth dorsal and fourth lumbar vertebræ The lesion when associated with neurologic manifestations shows a definite predilection for the mid thoracic area probably due to the fact that the spinal canal is narrowed at the mid thoracic level and the cord can be encroached upon more easily by an extramedullary lesion in this region

Pathogenesis Compression myelitis may be due to gradual collapse of the vertebral body This is rare and other factors are probably more significant In the cases associated with neurologic findings, the lesion affects not only the vertebral body but also shows a high incidence of involvement of the spinous processes, the laminæ and the pedicles *Compression of the spinal cord cannot be explained solely on the basis of* extensive involvement of a vertebra since cases in which the entire



Fig 312 Hemangioma of the Vertebra A Anteroposterior view B Lateral view The body of the third lumbar vertebra shows increased radiance with multiple fine trabeculations which are arranged in honeycomb fashion The changes extend into the lamina and the pedicles of the vertebra

vertebral *segment is affected by the tumor may present no signs* of cord compression Ballooning of the vertebra is frequent and may be the result of edema and thrombosis with resultant encroachment upon the spinal canal In rare cases there may be epidural hemangiomatous involvement

Clinical Manifestations Most cases of hemangioma of the vertebra present no clinical manifestations being discovered accidentally during roentgen study of the spine or at autopsy The symptoms are referable to the spinal cord, correspond to the level of the osseous lesion and vary from mild local pain and tenderness in the region of the spine to paraplegia and sphincteric incontinence The progression of the symptoms may be slow or rapid and remissions are frequent The extent and character of the neurologic manifestations depend upon the degree of spinal cord compression and/or damage The earlier findings are usually those referable to the dorsal and lateral columns of the spinal cord and are manifested by spasticity and paresis of the lower extremities associated with ataxia Spinal fluid examination is the only laboratory test of

diagnostic value With compression of the spinal cord, the spinal fluid may show xanthochromia and elevated protein levels A positive Queckenstedt sign can usually be demonstrated

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Tumors of the Mandible

The neoplasms of the jaw are best considered as a separate group for the reason that they present certain special characteristics not usually found in other regions of the body

Root Cysts These are usually residual from a previous apical abscess There is expansion and thinning of the cortex with multiple trabeculae in most instances

Dentigerous or Follicular Cysts These lesions develop from an un erupted tooth bud and may contain one or rarely several teeth There is erosion of bone with little or no new bone formation

Odontoma Odontomas arise from the pulp organ and are comprised of dentine, cementum and enamel The lesion occurs in the region of the root of the tooth and forms a dense, hard, bony mass There is expansion of the jaw with irregular, dense mottling, and in some instances a shell of dense bone around the neoplasm

Epulis (Benign Giant Cell Tumor) Epulis occurs more frequently in adults and usually is found at the margin of the mandible or maxilla There *is bone destruction about one or two teeth with extension through the* cortex producing a soft, circumscribed mass in the gum There may be



FIG 313 Root Cyst There is a rounded sharply defined area of increased radiance in the inferior first molar region The apices of the affected tooth are partially absorbed

trabeculation within the involved area otherwise the appearance is similar to dentigerous cyst The margins of the lesion are sharply defined

Adamantinoma The adamantinoma is an epithelial neoplasm of the basal cell type While usually benign the tumor not infrequently presents evidences of local malignancy with invasion of neighboring organs and regional or distant metastases The mandible is the most common site The tumor has also been reported in the pituitary body, the lip the cheek and the tibia Adamantinoma occurs at widely varying age periods the disease having been recorded in every decade of life The Negro race is apparently four times more frequently affected than other races Adamantinomas of the jaw may originate from several sources all of which derive from the parent cell the oral epithelium The principal sources comprise the oral surface epithelium the mucous glands of the oral epithelium epithelial cell inclusions along the teeth, the enamel organ of the primitive enamel anlage of a degenerated dental follicle, and follicular cysts In the jaw the condition is characterized by a long, slow course It is a painless centrally located tumor, hence produces no symptoms until it has attained considerable size A loose or painful tooth precedes the appearance of the growth It has been stated that there is

no record of these tumors occurring in a mouth with a full set of erupted teeth except in the presence of an impacted or otherwise malposed tooth. In some instances a fistula develops in the buccal cavity and there is discharge of straw colored or clear fluid. On palpation there may be crackling of the thin distended bony shell. Tumors originating in the maxilla usually invade the antrum and the nasal cavity.

Roentgen Manifestations. The roentgen manifestations are characteristic. The solid adamantinoma produces a monocystic shadow on the roentgenogram. It is differentiated from the odontogenic cyst by the appearance of the border of the bone defect. This is smooth and continuous in the cyst but in adamantinoma is interrupted by small niches with the production of a lobulated appearance and fine bone trabecula



Fig. 314. Adamantinoma. There is an irregular multilocular area of radiance in the body of the mandible. The mandible is widened.

at the anterior and posterior margins of the lesion. The polycystic type presents a honeycomb appearance in the early stage. Later there is huge distention of the bone by a cystic defect with multiple round compartments. The central giant cell tumor does not attain as large size as an adamatinoma. Chondromas produce fine straight bone trabeculation and do not form round cystic compartments. In some instances an adamantinoma forms a huge single cavity similar to a dentigerous cyst. Adamantinoma and dentigerous cyst may contain a tooth, although it is more common in the latter. The location of the tooth in the cyst serves as a diagnostic aid. In the dentigerous cyst the crown of the tooth is pushed away from the gingiva to the periphery of the cavity while in adamantinoma the tooth may be located in any portion of the lesion as it is imbedded and fixed in the tumor tissue. The lobulated polycystic appearance of the bone defect is characteristic and a definite diagnosis can be made on the basis of the roentgen findings.

MALIGNANT BONE TUMORS

SARCOMA OF BONE

This *group comprises all malignant neoplasms originating in bone* according to the Registry of Bone Sarcoma Others prefer to limit it to neoplasms arising from bone-producing cells However, it is generally agreed that osteogenic sarcoma is properly spoken of as malignant degeneration of bone producing cells The process usually originates within the bone, most frequently in or near the end of the shaft The growth extends into the cancellous portions of the bone and breaks through the cortex with resultant formation of a rounded or oval mass which tends to encircle the bone The new bone formed in the involved area varies according to its location Within the bone, it forms by direct metamorphosis of fibrous tumor or neoplastic osteoblasts into bone Bone tumor extrinsic to the cortex is formed by fibrous or endochondral ossification The process is characterized by extensive bone destruction occurring simultaneously with bone proliferation While osteogenic sarcoma may occur at any age, it is most common in young adults of the second decade of life Males are more often affected than females The long bones are most frequently involved, and in the following order femur tibia humerus and fibula Metastases are very common The lung is the usual site Other bones and the lymph nodes may also be involved The metastases usually disseminate by way of the blood stream although the lymphatics may also serve to spread the process The distant lesions are usually similar to the primary lesion morphologically

Pathogenesis There are two types of sarcoma of bone osteogenic sarcoma and fibrosarcoma Periosteal fibrosarcoma originates from the connective tissue of the outer layer of the periosteum The tumor is well differentiated rich in fibrous elements, firm and well defined The lesion grows slowly and only rarely causes destruction of the neighboring bone All other sarcomas of the supporting tissues of the skeleton are classified as osteogenic sarcoma This term is often interpreted erroneously as meaning bone-forming sarcoma It denotes a sarcoma of the osteogenic or loose connective tissues the cells of which may differentiate into osteoblasts The term "osteogenic sarcoma actually is not appropriate because tissue normally differentiating into cartilage may also give rise to similar tumors However it is so well established and widely used that change in the nomenclature would cause further confusion and is inadvisable

*To understand the biological meaning of the terms osteogenic and chon*drogenic tissue it is essential to understand the peculiarities of growth of cartilage and bone Cartilage grows interstitially and by apposition At the margins of the articular cartilage growth is by extension of the synovial membrane in the region of its insertion to bone Appositional growth results in widening of the articular cartilage Epiphyseal cartilaginous plates grow in transverse diameter by apposition at their perichondral surface Bone grows by apposition In interstitial growth the specfic and differentiated tissue cells produce cells only on their own level of differentiation In the growth of cartilage by apposition the cells of the surrounding connective tissue termed the perichondrium differentiate into chondrocytes the process repeating itself as long as the cartilage

grows The cells of the periosteum, the endosteum, or both differentiate into osteoblasts New osteoblasts must differentiate after the disappearance of osteoblasts in periods of rest or after resorption necessitating the presence of pluripotential cells in the connective tissue which surrounds growing bone and cartilage Connective tissue in any part of the body may have a specific chondrogenic and osteogenic property These properties are not limited to periosteum and endosteum In order to initiate the production of bone, multiple factors are necessary, an important one being a localized concentration of phosphatase Other stimuli of chemical or mechanical nature are also essential to initiate bone production The bones are surrounded by and contain pluripotential mesodermal cells and these elements may differentiate into chondroblasts osteoblasts and fibroblasts The cells are not restricted to any part of the skeleton or particular period of life as the continuous reconstruction of bone requires apposition of new bone tissue after resorption of overaged bone or bone that is mechanically inadequate

In a malignant tumor the growth and multiplication of cells is pathologically exaggerated The cells of the original tissue are dedifferentiated to a very low level and the structure and potencies are greatly distorted In the presence of a tumor of mesodermal origin, the cells may again produce intercellular elements The resulting tissue can in many instances not be identified with a normal tissue and in most cases the tissue of the tumor shows only certain similarities to a normal tissue No tumor other than osteogenic sarcoma shows such wide variation in histologic structure Only rarely do two osteogenic sarcomas have the same structure, and it is impossible to find an osteogenic sarcoma with but one type of tissue throughout its entire extent Different areas of the same tumor may show marked differences in structure This is due to the fact that osteogenic sarcoma originates from pluripotential cells of relatively low differentiation The sarcomas consist of connective tissue cartilage and bone and each of these tissues is present in many variations

When bone is formed in an osteogenic sarcoma it presents a widely variable histologic picture Mature lamellated bone is not produced islands of mature bone in the tumor representing merely remnants of old bone The tumor itself does not resorb bone The growing tumor can only stimulate the normal connective tissue to differentiate osteoclasts The resorption of bone ceases as the connective tissue separating the tumor from bone is destroyed by the growing tumor and the tumor cells come in direct contact with the bone In osteogenic sarcoma the gross and microscopic findings and the roentgenographic pictures differ according to the amounts of bone formed With large amounts of dense bone, the tumor is termed sclerosing or osteoblastic, whereas if bone destruction prevails the tumor is termed osteoclastic Destruction of bone appears to be common in all osteogenic sarcomas The vascularization of an osteogenic tumor is variable In certain tumors there is an abundance of large thin walled blood vessels and capillaries and the tumors are referred to as telangiectatic sarcoma or malignant bone aneurysm These are more malignant than other types of osteogenic sarcoma as invasion of blood vessels by tumor cells with development of metastases occurs early Hemorrhage into the tumor or necrosis of portions of the tumor may lead to the formation of cystic cavities In osteogenic sarcoma metastasis is most often to the lungs because of invasion of the veins by tumor cells The cells of the tumor may produce in the metastatic focus cartilage or bone similar to the tissue of the primary tumor

General Characteristics While the classification of malignant tumors of bone is of great value, roentgen methods of study do not always make it possible to differentiate osteogenic sarcoma into its several varieties Whether medullary, subperiosteal telangiectatic or sclerosing, there is always an admixture of osteolytic and osteoblastic elements involving the medulla, cortex and periosteum in varying degrees The first sign may be localized osteoporosis of the medulla, cortex or both This is followed



Fig 315 Fig 316

Fig 315 Osteogenic Sarcoma There are numerous ray formations extending at right angles to the shaft of the lower third of the femur The periosteum is elevated and there is a large soft tissue mass adjacent to the bone

Fig 316 Osteogenic Sarcoma of the Femur There are destructive and proliferative changes involving the lower third of the shaft of the femur The new bone formation is in the form of multiple spicules radiating at right angles to the shaft in characteristic sun ray formation There is elevation of the periosteum along the anterior aspect of the lower end of the femur The soft tissues about the lower end of the femur present a large tumor formation

by extensive medullary destruction Where the cortex is involved the tumor may extend beyond it into the soft tissue leaving ragged remnants of the original cortex in place New bone formation is manifested by thin bands of calcification adjacent to the shaft under the raised periosteum "Sunray spicules may be produced by new bone formation at right angles to the surface of the bone under the raised periosteum and are found in 18 per cent of these tumors as well as in certain cases of chronic osteomyelitis and syphilitic periostitis The reactive triangle of Codman is shown on the x ray as a triangle of subperiosteal new bone merging into

the surface of the bone above and below the tumor and with an ill defined edge Between these limits the subperiosteal new bone is destroyed by the advancing tumor While osteolytic or osteoblastic activity may predominate in the tumor there is always evidence of both bone destruction and bone production Eventually the periosteum is perforated and the tumor extends into the soft tissues with the formation of a mass

The medullary, subperiosteal sarcomas arise from the shaft and extend both underneath the periosteum and into the marrow cavity The neoplasm may grow very rapidly or slowly, the two varieties having different characteristics The rapidly growing type is composed of small round cells and sweeps everything before it with extensive destruction of the cortex, there being no time for bone atrophy or expansion of the cortex The lesion is manifested by an area of destruction in the central portion of the shaft usually near the end of a long bone One side of the shaft may be completely destroyed with invasion of the soft parts There is no periosteal elevation or proliferation and no new bone formation is present in the soft tissue In the slow growing type, there is expansion of the cortex with little erosion until late in the course of the disease The changes closely simulate those in bone cyst or other benign bone lesions such as enchondroma giant cell tumor or osteomyelitis

Telengiectatic osteogenic sarcoma is a form of malignant bone aneurysm Its exact origin and relation to other bone sarcomata have not been established The lesion is usually circumscribed Some are entirely destructive while others form new bone in varying amounts Periosteal sarcoma is a highly malignant rapidly growing neoplasm It is of spindle cell structure originates from the outer layers of the periosteum and is characterized by preservation of the shaft Trauma may precede the onset of the neoplasm Rapid growth and early metastases usually occur The metastases may be in the lungs brain, lymph nodes and the periosteum of other bones There are sunray or needle like radiations of new bone at right angles to the shaft extending for various distances into the soft tissues The shaft of the bone presents little or no bone destruction and there is no expansion of the cortex A large soft tissue mass is palpable and can be demonstrated roentgenographically Parosteal or capsular osteogenic sarcomas arise from the deep fascia and fibrous capsule of the joints, not from true bone tissue They often produce bone and cartilage and must be differentiated from fibrocellular synoviomas and chondromas

Sclerosing Osteogenic Sarcoma Osteosarcoma

Osteosarcoma occurs at all ages from childhood to advanced age In many instances the lesion terminates fatally in a few months in others the disease persists for as long as twenty-five to thirty years The tumor is characterized by extensive destruction of bone which is irregularly mottled in character The process may be osteolytic or associated with new bone formation and reactive periostitis of varying degrees Extensive calcification may occur within the bone or in the extra osseous portions of the tumor, the new bone formation being manifested by small or large calcific areas within the osteolytic zones formed by the tumor The tumor mass in the soft tissues may show extensive or moderate calcification The periosteum is elevated and there is a sharp angulation at the point of separation of the periosteum from the bone, the so called periosteal

General Characteristics While the classification of malignant tumors of bone is of great value, roentgen methods of study do not always make it possible to differentiate osteogenic sarcoma into its several varieties Whether medullary subperiosteal, telangiectatic, or sclerosing there is always an admixture of osteolytic and osteoblastic elements involving the medulla cortex and periosteum in varying degrees The first sign may be localized osteoporosis of the medulla, cortex or both This is followed



Fig 315 Fig 316

Fig 315 Osteogenic Sarcoma There are numerous ray formations extending at right angles to the shaft of the lower third of the femur The periosteum is elevated and there is a large soft tissue mass adjacent to the bone

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Fig 317 Fibrosarcoma of Femur *A* Anteroposterior view *B* Lateral view There is a localized area of bone destruction in the lower end of the shaft of the femur The cortex of the anterior aspect of the lower femur shows a localized area of destruction with elevation of the periosteum The patient was a female thirty seven years of age For about four months there had been severe pain and a large soft tissue mass had developed in the region above the knee Amputation was performed and histopathologic study of the specimen confirmed the roentgen diagnosis of fibrosarcoma

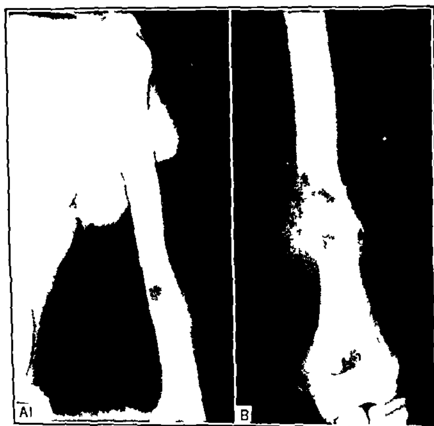


Fig 318 Fibrosarcoma of the Humerus *A* There is an area of increased radiance in the region of the junction of the lower and middle thirds of the shaft of the humerus The margins of the area are irregular in outline and there are numerous fine bony trabeculations within it The cortex is thinned and there is a pathologic fracture *B* Two months later The process has advanced markedly Amputation was performed Pathologic report fibrosarcoma

lipping or Codman's triangle The so called sunray or sunburst formation in the tissues which is the characteristic manifestation of osteosarcoma is present in many instances and may constitute the only roentgen manifestation of the neoplasm in some instances The lesion is most commonly seen in the diaphyses or metaphyses and frequently extends beyond the epiphyseal line Extension into the soft tissues with formation of an extra osseous mass may ensue

Osteolytic Sarcoma Osteolytic Osteogenic Sarcoma

Osteolytic sarcoma is a highly malignant tumor, many patients with this lesion succumbing in three or four months However, many live for five or more years The disease has been found at all ages from six years to sixty or more years It occurs in the metaphysis as does giant cell tumor and is confined to the metaphyses in the early stages and prior to the closure of the epiphyseal plate The lesion is much more extensive than the roentgen changes indicate

Roentgenographically, osteolytic sarcoma is characterized by an irregular, poorly defined area of mottled rarefaction in the bone Marginal sclerosis is absent Periosteal reaction is usually present and is extensive The tumor is invasive and there is both central and subperiosteal involvement Portions of the shaft remain unaffected Osteoblastic changes are less pronounced than in the sclerosing type of osteogenic sarcoma There is usually a large soft tissue mass adjacent to the bone lesion The mass in many cases is much more extensive than the bone lesion

Fibrosarcoma

Fibrosarcoma is purely osteolytic in character The tumor may originate in the bone or the soft tissues It may occur at any age from childhood to the seventies the average being about thirty five years In some instances it results fatally in a few months while others persist for ten, fifteen or more years The lesion may arise in a scar subsequent to inflammation

Roentgen Manifestations There is an area of localized destruction in the bone The margins of the lesion may show increased density due to reactive sclerosis Periosteal reaction is slight The bone destruction is irregular and moth eaten in character, although a sharply defined, uniform area of destruction may occur The roentgen appearance may suggest a benign process or the presence of a previous benign lesion such as fibrous dysplasia or xanthoma A large soft tissue mass forms after the lesion has extended through the cortex A connection with Paget s disease has been suggested because of the frequent coincidence of these two lesions

Chondrosarcoma

Chondrosarcomas arise from pre existing normal cartilage, remnants of cartilage displaced by rickets chondromas or ecchondroses They tend to be bulky and may become very large The lesion may be central or peripheral and tends to calcify and ossify irregularly They grow slowly and metastasize late The upper femur and ribs are the commonest sites The chondrosarcomas occur in the age groups from ten years to seventy five years, the average age being about twenty five to twenty six years The duration of the reported cases has varied from two months to



Fig 319 Chondrosarcoma of the Femur *A* There is extensive destruction with *irregular* mottling involving the upper third of the shaft *of the* femur A large rounded soft tissue mass lies below *the neck* and medial to the upper end of the shaft of the femur The lesser *trochanter* is separated There is marked osteoporosis of the head and neck of the femur *B* Six weeks *later* after the administration of deep *x* ray therapy There is extensive recalcification of the lesion with increased bony density *throughout* the involved area and within the soft tissue mass The soft tissue mass has decreased *markedly in size*

fourteen years, the average is two and six tenths years Chondrosarcoma is a tumor which produces cartilage The cartilage frequently undergoes calcification and the lesion in these instances is characterized by flocculent calcific depositions which are manifested roentgenographically as faintly visualized or marked areas of increased density The calcific shadows are distributed in irregular fashion throughout the neoplasm, *usually being most marked in the extra osseous portions of the tumor* Chondrosarcomas which are uncalcified may attain considerable size yet give little or no roentgen evidence of their presence, a slight patchy reaction in the bone being the sole manifestation This is particularly apt to occur in the bones of the pelvis Periosteal chondrosarcomas are extremely rare These chondrosarcomas are associated with solid calcification and in some instances perpendicular ray formations which are closely similar to those in osteosarcomas

Chondrosarcomas are divided into two groups, the central and the peripheral The manifestations of the central type vary according to whether the lesion originates in the medulla or in the cortex under the periosteum Those which begin in the medulla show a large, thick-walled cavity or cavities which tend to destroy the cortex The areas of decreased density may show trabeculation with central areas of multi locular destruction or foci of calcification scattered irregularly throughout the lesion They occur in the ends of the long bones The cortex is not perforated until relatively late The peripheral type, which originates in the cortex directly under the periosteum, shows a faintly visible shadow in the soft tissue adjacent to the bone The periosteum is elevated In many instances there is no medullary or cortical involvement The roentgen changes may be very slight Calcification is sparse and takes the form of radiating spicules at right angles to the cortex or large flecks The lesion may cast so faint a shadow on the roentgenogram that the diagnosis is not established The perpendicular spicules which occur in this tumor differ from those of osteogenic sarcoma in that the former grow to greater length and sometimes measure two or more cm while in osteogenic sarcoma they usually do not measure over one or two cm in length The radiating spicules in chondrosarcoma present a flat appearance suggesting that the peripheral surfaces of the spicules are molded by the overlying muscles Secondary or peripheral chondrosarcoma developing on the basis of benign osteochondroma usually retains the salient features of the primary tumor The edges become ragged and there is a large soft tissue mass with irregular foci of calcification

Malignant Osteoclastoma Malignant Giant Cell Tumor

The osteoclastoma is usually benign but it may be malignant In many instances it appears that the tumor is malignant from the onset In this type of case death occurs from pulmonary metastases Usually the lungs are the only site of metastatic involvement but in some cases the metastases are widespread deposits being found in the skin subcutaneous tissues, bone lymph glands and other parts of the body A malignant osteoclastoma may be termed an osteogenic sarcoma as a sarcoma is any malignant tumor derived from the connective tissues However this terminology is inaccurate If an osteoclastoma is malignant or becomes malignant it still shows sufficient differentiation towards the formation of osteoclasts to determine its type

the new bone is dense and solid and closely simulates osteomyelitis of the proliferative type The tumor growth may be so rapid that the new bone grows outward from the shaft of the involved bone in the form of a dense mass or rays, especially in the regions of the ends of the neoplasm The shafts of the long bones may show a fusiform enlargement with parallel layers of bone involving almost the entire length of the shaft The peripheral layers of new bone may appear to end free in the soft tissues Bone destruction is usually not present, nor is there expansion of the bone At the upper and lower margins of the tumor, there is a triangular elevation of the periosteum with areas of new bone formation which are dense and lamellated Metastases are common and are usually by way



Fig 321 Ewing s Tumor There is irregularly increased density with scattered areas of rarefaction involving the neck and upper portion of the shaft *of the femur* Small bony spicules extend irregularly into the soft tissues in the region of the inferior aspect of the neck and about the lesser trochanter Biopsy report Ewing s tumor

of the blood stream The lungs are the commonest site Other bones may be involved

The *first roentgen* manifestation is usually an area of osteoporosis This is diffuse and progresses to an extensive area of bone destruction There may be localized sclerotic changes although in other instances patchy or diffuse reactive bone formation occurs Ewing's tumor may not produce any dissolution of continuity of the outline of the cortex It is frequently characterized by an elliptic expansion of the periosteum of the shaft of the long bone In those cases in which the tumor breaks through the cortex, there is abrupt dissolution of the continuity of the contour The laminated appearance of the cortex and the break in continuity may occur simultaneously As the tumor invades the contiguous soft tissue

1 Ewing's Sarcoma, Endothelioma, Hemangioendothelioma
2 Angiosarcoma

In this group are included both endothelioma (Ewing's tumor) and angiosarcoma as these lesions produce similar manifestations Ewing states that the typical endothelioma of bone arises from capillary endothelium and never exhibits any properties other than those belonging to vascular endothelium It is the only malignant tumor which arises from the cortex of bone Ewing's tumor may occur at any age, being most

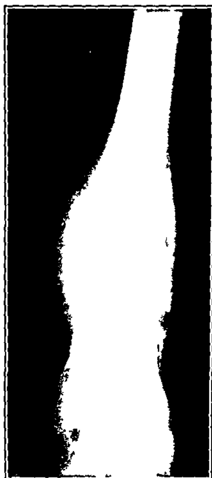


Fig 320 Ewing s Tumor There is extensive rarefaction involving the lower third of the shaft of the femur There is marked elevation of the periosteum with multiple bony spicules extending into the soft tissues adjacent to the shaft of the femur

frequent in the second and third decades and may occur in practically any bone The most common site is the region of the midshaft of the long bones It spreads into the medullary canal infiltrates the cortex and frequently lifts the periosteum New bone formation occurs along the outer aspect of the cortex and also within the cancellous spaces in the region of the growth The neoplasm may break down at its deeper portions while continuing to proliferate at the edges It characteristically tends to extend through successive layers of newly laid down periosteal bone with the formation of parallel layers of bone one above the other producing the so called onion skin effect along the shafts of the long bones the most typical manifestation of the disease In other instances,

the new bone is dense and solid and closely simulates osteomyelitis of the proliferative type. The tumor growth may be so rapid that the new bone grows outward from the shaft of the involved bone in the form of a dense mass or rays, especially in the regions of the ends of the neoplasm. The shafts of the long bones may show a fusiform enlargement with parallel layers of bone involving almost the entire length of the shaft. The peripheral layers of new bone may appear to end free in the soft tissues. Bone destruction is usually not present nor is there expansion of the bone. At the upper and lower margins of the tumor there is a triangular elevation of the periosteum with areas of new bone formation which are dense and laminated. Metastases are common and are usually by way

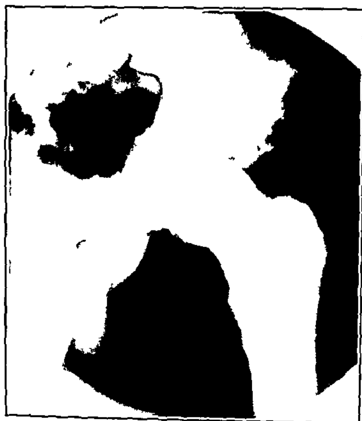


FIG 321 Ewing's Tumor *There is irregularly increased density with scattered areas of rarefaction involving the neck and upper portion of the shaft of the femur. Small bony spicules extend irregularly into the soft tissues in the region of the inferior aspect of the neck and about the lesser trochanter. Biopsy report Ewing's tumor*

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The first roentgen manifestation is usually an area of osteoporosis. This is diffuse and progresses to an extensive area of bone destruction. There may be localized sclerotic changes, although in other instances patchy or diffuse reactive bone formation occurs. Ewing's tumor may not produce any dissolution of continuity of the outline of the cortex. It is frequently characterized by an elliptic expansion of the periosteum of the shaft of the long bone. In those cases in which the tumor breaks through the cortex there is abrupt dissolution of the continuity of the contour. The laminated appearance of the cortex and the break in continuity may occur simultaneously. As the tumor invades the contiguous soft tissue

structures, the margins of the invading shadow are not as clearly demarcated as in a benign tumor, yet they can be discerned faintly in contradistinction to the promiscuous invasion which occurs in the osteogenic sarcoma group. While the disease is progressive and rapidly fatal in some instances, other cases present a localized tumor which persists for many years prior to the development of metastases. In many instances Ewing's tumor responds favorably to radiotherapy. This may be an important aid in diagnosis. After treatment there is marked ossification subperiosteally and the periosteal shell becomes thickened and sclerotic producing an appearance similar to that of the involucrum in healed osteomyelitis. Permanent cures have been reported after irradiation. Ewing's tumor simulates osteomyelitis very closely and differentiation from pyogenic bone lesions is frequently very difficult.

MULTIPLE MYELOMA

The exact nature of multiple myeloma and the origin of the predominant cell of the disease have not been established with definiteness. Ewing considers it a specific malignant tumor which arises from a single cell type. Others are of the opinion that the myelomas are made up of cells of the same fundamental types in various stages of development, differentiation, and degeneration. The cell is described as plasma like with an eccentric nucleus containing a spoke-like arrangement of chromatin. The cells have been termed *lymphocytes, lymphoblasts, myelocytes, myeloblasts,* erythroblasts, and plasma cells. The best terminology is to refer to all members of the group as myeloma cells. It is relatively uncommon but not rare. The various forms may resemble carcinoma, leukemia, and lymphosarcoma. It should be regarded as a neoplastic disease which occupies a place between the frank tumors and the leukemias. The usual site of origin is the bone marrow, although extramedullary origin may be encountered. The disease may present itself in single or multiple foci. It tends to involve principally the ribs, spine, skull and pelvis. The disease is uniformly malignant and fatal. The average duration of life is two and one half years after the diagnosis has been made. Bone changes are not demonstrable in about 60 per cent or more of the cases. The disease is referred to in the literature as plasmacytoma, plasmoma, erythroblastoma, myelocytoma, plasma cell myeloma, and plasmacytic leukemia.

Clinical Manifestations. Multiple myeloma has been recorded from the second to the eighth decade of life, the average being about fifty five years. There is no characteristic clinical picture. Pain is the most common feature and may be mild or severe. Fractures are frequent and the incidence of pathologic fractures may be relatively high, in one series being recorded as 62 per cent of the patients with rib involvement. Palpable tumors are present in many cases. There is albumin and Bence Jones protein in the urine. The albumin globulin ratio is reversed. The blood calcium is elevated and the serum phosphorus is normal or nearly so.

Roentgen Manifestations. The roentgen manifestations are widely variable. In many instances there are no demonstrable bone changes. In others there is a diffuse generalized demineralization which cannot be distinguished from senile or postmenopausal types of osteoporosis. This is frequently accompanied by compressions of the vertebral bodies. In some cases the demineralization is spotty and produces a granular or stippled effect. This is particularly apt to occur in the ribs, the skull,

and the pelvis A honeycomb appearance in the trabecular structure of the proximal portions of the shafts of the femurs and humerus may be present the latter effect being accentuated against the background of the radiolucent medulla Osteolytic lesions are characteristic of the disease and vary widely in number size location and structural appearance In the solitary or isolated lesions there is a cystic trabeculated appearance which closely simulates giant cell tumor There may be considerable expansion associated with the destruction and the process may resemble osteochondrosarcoma The solitary lesion may persist for weeks or months The most typical form of the disease in the bone is the wide spread sharply demarcated, oval or rounded, punched out areas of in creased radiance which establish the diagnosis with definiteness The defects occur in the skull ribs pelvis and other bones and closely resemble osteolytic metastases of carcinoma, especially of the breast The previously accepted concept that the "punched out defect was the uni

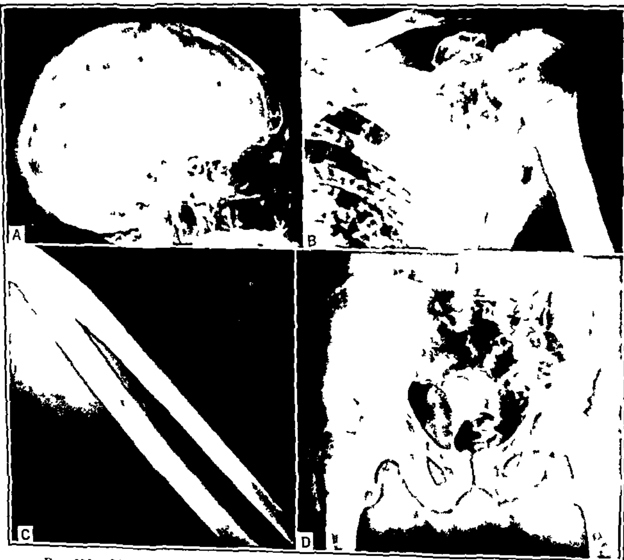


Fig 322 Multiple Myeloma *A* Skull lateral view *B* Shoulder *C* Forearm *D* Pelvis and Hips There are multiple rounded areas of increased radiance in the skull ribs clavicles scapulas femurs and bones of the pelvis *the characteristic manifestation of multiple myeloma* The scapula and left ilium show large *cystic areas* which comprise a less common manifestation of the disease these cyst like changes are easily *confused* with giant cell tumor or echinococcus disease

versal bone change has been discarded and it is now generally appreciated that the classical feature of multiple discrete areas of destruction is the exception rather than the rule Lesions in the spongiosa are more easily detectable than those in the cortex, defects measuring 8 to 10 mm in diameter in the cortex of a long bone or a vertebra being invisible while similar areas of destruction may be demonstrable in the spongiosa

The localized area of bone destruction is most common in the skull This is due to the thinness of the bone and the relatively small amounts of overlying soft tissue permitting of readier detection of small, isolated lesions The areas of destruction are visualized as sharply defined zones measuring 2 mm to 1 cm in diameter The defects occur in all portions of the skull and may closely resemble pacchionian bodies or diploic venous lakes Coalescence of smaller lesions results in large areas of destruction of bone The skull may remain uninvolved in the presence of extensive disease elsewhere in the skeleton The most common change in the spine is osteoporosis There may be thinning of the trabeculæ and extensive obliteration of the architecture of the bone only the cortex remaining The presence of multiple bone lesions without involvement of the spine militates against the diagnosis of myeloma The lower dorsal and lumbar spine are most apt to be involved The vertebræ are frequently collapsed and wedge shaped While multiple involvements are common contiguous vertebræ are rarely affected Soft tissue masses are frequently present in the region adjacent to the bone lesions as in tuberculosis and lymphoma Extension across and around the intervertebral disc is rare but may occur A bizarre finding in multiple myeloma is proliferative new bone formation involving the humeral shaft Usually osteosclerosis occurs in myeloma only after fracture or subsequent to roentgen therapy

Pathogenesis Multiple myeloma is a distinctive malignant disease of the skeleton It originates in the myeloid formative tissues Practically every bone in the body may be involved It is important in diagnosis to stress the significance of hypercalcemia, hyperglobulinemia and Bence Jones proteinuria The albumin globulin ratio is reversed in about 50 per cent of the cases There may be atypical amyloidosis and distinctive cytologic changes in the kidney in association with the myeloma There are definitely authenticated cases in infants and children between the ages of sixteen months and twelve years

The two lesions which fall into this group comprise the myeloma and the reticulocytoma Lipoma and liposarcoma of the skeleton may rarely originate in the adipose tissue of the bone marrow and may also be included in this group There are stem cells of erythrocytes The granulocytes are identical primary or stem cells and in the early stages of their development have more than one developmental potentiality Tumors may arise from any of these cells of low differentiation The cells may be similar to those of the later stages in the development of the blood cells The independently growing and differentiating cells of the tumor may assume a morphologic character which is not present in normal tissues These manifestations indicate the degree of variability in the myeloma group In the past the tumors were referred to as erythroblastoma lymphocytoma plasmacytoma and myelocytoma These terms should be abolished or utilized only with the understanding that they indicate merely a superficial similarity to the tumor cells of certain elements of the bone marrow The use of the term myelocytoma for all varieties of

myeloma is preferable for the reason that it indicates the common source of the tumor cells Another source of tumors of the bone marrow is the supporting or reticular connective tissue with its characteristic cells termed the reticulocytes This type of tumor is called the endothelioma or reticulocytoma

On the basis of the type of cell the following classification is commonly utilized (1) myelocytoma, the cells being similar to the myelocytes of the normal bone marrow (2) erythroblastoma the cells containing hemoglobin, (3) lymphocytoma consisting of lymphatic tissue comparable to that in lymphosarcoma, and (4) plasmacytoma the cells showing eccentric nuclei and vacuoles typical of the plasma cell



FIG 323 Multiple Myeloma There are extensive destructive changes in the ribs scapulas and clavicles The lesions are osteolytic in character with no evidences of new bone formation The roentgen diagnosis of multiple myeloma was confirmed by histopathologic study

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portions. On microscopic examination the appearance is similar to reticulum cell sarcoma of the lymph nodes, the viscera and other portions of the body. The cells are larger than normal lymphocytes, are round, oval or reniform and vary widely in size and shape. The tumor as it grows destroys and replaces the normal architecture of the bone marrow with destruction of the cortex and marked new bone formation. Studies of the chemistry of the blood reveal no significant abnormalities. Anemia is not an important factor. The white cell count is not elevated. This may be of value in ruling out osteomyelitis.



FIG. 324. Reticulum Cell Sarcoma of the Right Femur. *There is extensive mottling with multiple areas of rarefaction in the lower third of the shaft of the femur. There is slight irregularity of outline in this region. There is no periosteal elevation and no soft tissue mass is demonstrable. The knee joint space is not involved. There is marked osteoporosis of the bones about the knee. Biopsy report: reticulum cell sarcoma.*

Symptoms. The clinical manifestations are similar to those of other malignant tumors arising in bone. Pain is relatively frequent and is usually localized to the involved bone or may be referred to the nearest joint. Swelling is present in a large number of cases and involves the soft tissues contiguous to the tumor. In many instances the soft tissue swelling is larger than the tumor in the bone and may reach enormous proportions. A striking feature is that the patients remain in a state of general well being even in the face of advanced disease. The symptoms frequently are of long duration prior to the discovery of the lesion. On physical examination there is usually a palpable tumor, edema of the overlying

The solitary form of myeloma is very rare and is most probably not a separate entity, rather an early phase of the disease Most cases of solitary lesion remain localized for a long period of time The solitary myeloma may occur in a cystic form or as a trabeculated lesion This type may be purely destructive and closely simulate metastatic carcinoma The distinction between the solitary and generalized forms of myeloma is artificial as the existence of a solitary lesion merely represents an early stage of the same disease It appears probable that multiple myeloma begins as an unrecognized solitary myeloma and when demonstrable roentgenographically, the disease has become generalized

Differential Diagnosis In differential diagnosis one must consider giant cell tumor eosinophilic granuloma Ewing s sarcoma osteolytic metastases, and Paget s disease In metastatic carcinoma vertebral collapse occurs late and the discs are not affected Differentiation from tuberculosis of the spine is difficult as this disease presents closely similar changes The absence of a paravertebral abscess and the lesions in the other parts of the skeleton establish the diagnosis of myelomatosis In xanthomatosis, the deficiencies in the skull may simulate the lesions of multiple myeloma Xanthomatosis is more common in children and young adults and the areas of rarefaction in the skull tend to assume a map like configuration Osteomalacia and osteitis fibrosa cystica must also be considered in differential diagnosis Paget s disease and multiple myeloma are common While the co existence of these diseases is very unusual both occur in the same age period and they may exist simultaneously The changes in multiple myeloma can easily be confused with those in Paget s disease The chief point of differentiation may be in the skull There is lack of increase of thickness of the skull in multiple myeloma An important factor in the diagnosis of multiple myeloma is the occurrence of abnormality of the plasma proteins The diagnosis may be aided by a careful study of the serum calcium and inorganic phosphorus levels and a determination of the alkaline and acid phosphatase activity

PRIMARY RETICULUM-CELL SARCOMA OF BONE

Primary reticulum cell sarcoma of bone is a malignant tumor which is histologically identical with reticulum cell sarcomas in other regions of the body The neoplasm arises in a single focus in the bone It is malignant and is capable of regional and distant metastases The disease is most common in the earlier decades of life The tumor is slow growing and pursues a relatively long course It is sensitive to radiation and in many instances is curable by radiation There is usually no marked evidence of general debility It is a rare primary bone tumor The incidence is slightly greater in males than in females The lesion is most common in patients under forty the average age being about thirty one years Cases have been reported in individuals from eleven years to seventy years The disease is most common in the long bones but occurs also in the flat bones and in the vertebræ

Pathology The tumor is soft pliable and glistening While frequently homogeneous there may be areas of necrosis There frequently is erosion of the marrow and the cortex of the bone Although a pseudo capsule may be present the line of demarcation between tumor and the uninvolved segment of bone is usually poorly defined The neoplasm tends to extend into the adjacent soft tissues and may assume considerable pro

METASTATIC CARCINOMA OF THE SKELETON

Certain generalizations with respect to the development of bone metastases have been established The higher the grade of malignancy the younger the individual and the greater the extent of glandular metastases the earlier metastatic bone lesions usually occur The roentgen appearance of the metastatic lesions in the bone is not dependent on the degree of malignancy A more favorable response to therapy as regards the duration of the effect is usual in tumors of lower grades of malignancy The less malignant the tumor, the longer the duration of life after the development of metastases The course of the bone metastases in carcinoma is in general similar to the course of the primary neoplasm In cases of latent or asymptomatic primary carcinoma manifested by metastatic lesions, roentgen study is of inestimable assistance in the search for the primary site A thorough knowledge of the common and usual sites of metastases from the various types of malignancies is essential Abrams analyzed 1000 consecutive cases of malignant neoplasms of epithelial origin autopsied between 1943 and 1947 at the Montefiore Hospital and found that 27 2 per cent had metastasized to bone The skeletal metastases occurred in the following order of frequency

	Per Cent of the Cases		*Per Cent of the Cases*
Prostate	84	Rectum	12 5
Breast	73	Stomach	11
Thyroid	50	Ovary	9
Lung	32 5	Colon	9
Bladder	26	Esophagus	12
Kidney	23 5	Tongue	9
Uterus	22	Larynx	7
Pancreas	15 5	Cervix	7 5

Metastatic lesions spread through the blood and lymph streams the former being the more common route Involvement below the elbow and knee is rare although numerous exceptions occur Lymph borne metastases are usually to the bone adjacent to the primary lesion The most common examples of this are metastatic involvement of the bones of the pelvis femurs and spine in carcinoma of the prostate and of the ribs, spine and skull in carcinoma of the breast The bones most frequently involved in metastatic carcinoma are the ribs skull, vertebrae pelvis, femurs and humerus Certain malignant diseases apparently have predilection for particular bones Carcinoma of the thyroid is most apt to spread to the ribs Hypernephroma spreads to the skull, ribs and long bones the metastases are usually of the osteolytic type, single or multiple often attain large size and may give clinical evidence of their presence before the primary tumor many of which are asymptomatic for long periods of time Ewing s tumor is the bone neoplasm which most commonly metastasizes to other bones New bone formation may occur adjacent to the shaft of the bone and is parallel to the shaft or in the form of striations radiating from the bone and tends to be similar in appearance to the primary lesion Bone lesions not infrequently result from direct extension of neoplasms in the adjacent tissue Hypophyseal tumors, derived from Rathke s pouch tend to invade the skeleton Nasal polyps may

tissues, tenderness, loss of function, and atrophy The mass associated with the tumor in the case of the shaft of the long bones is most commonly fusiform In the flat bones or near a joint the tumor is apt to be ovoid and may be soft or hard and irregular In some instances the neoplasm appears attached to the underlying bone while in others it appears mobile Tenderness is a prominent feature and is localized in the region of the swelling There is no local heat or redness Temperature is usually not present Pathologic fracture may occur but is not a prominent feature of the disease The fractures heal after adequate x ray therapy The disease may prove fatal within eight months to five years, the usual duration of life being slightly over two years

Roentgen Manifestations The roentgen picture is not definitely characteristic in all instances The tumor appears to arise in the medullary portion of the bone It is predominantly destructive and osteolytic, production of bone being less prominent The areas of production are distributed in an irregular fashion and are interspersed with areas of destruction The cortex is destroyed without expansion Rarely the process is entirely destructive in character Periosteal thickening may be present with the production of dense new bone The direction of growth of the tumor is within the bone predominantly There is extension into the soft tissues, in some cases the extraosseous mass exceeding in size the area of bone involvement An important feature is the poorly defined boundary of the tumor both in the bone and soft tissues There is no characteristic location which is of diagnostic significance as the tumor may occur in any portion of the shaft and frequently crosses the epiphyseal line When the epiphysis is involved the lesion is usually not confined to that area Tumors in the region of the joint are frequently associated with synovitis Prompt response to roentgen radiation is a prominent feature of the disease and healing occurs with disappearance of the soft tissue mass re establishment of the cortex, and extensive new bone formation at the site of the tumor

Differential Diagnosis *The most difficult problem in differential diagnosis is osteogenic sarcoma* In osteogenic sarcoma there is extensive periosteal thickening and calcification of periosseous tumor masses There is a lesser degree of cortical destruction and medullary growth Osteogenic sarcomas are much less radiosensitive than reticulum cell sarcomas Ewing's sarcoma is difficult to differentiate as both conditions have many characteristics in common In Ewing's tumor the lesion occurs in the younger age groups is more rapidly progressive, and there are early and extensive pulmonary metastases There is more periosteal reaction in Ewing s tumor than in the reticulum cell sarcomas The simultaneous presence of bone destruction and proliferation within the bone may closely simulate osteomyelitis Differentiation can usually be made by the presence of fever response to antibiotics and the presence of bacterial growth on cultures Carcinoma metastasis to bone may occasionally simulate reticulum cell sarcoma In metastatic carcinoma of bone the cortical destruction is less marked there is less patchy destruction and no new bone formation The discovery of the primary source usually establishes the diagnosis Syphilis may cause confusion in diagnosis because of the periosteal reaction Other conditions which have to be considered in differential diagnosis include eosinophilic granuloma, osteoid osteoma, synovioma neuroblastoma angiosarcoma, malignant giant cell tumor myeloma Paget s disease bursitis synovitis and arthritis

METASTATIC CARCINOMA OF THE SKELETON

Certain generalizations with respect to the development of bone metastases have been established. The higher the grade of malignancy, the younger the individual and the greater the extent of glandular metastases, the earlier metastatic bone lesions usually occur. The roentgen appearance of the metastatic lesions in the bone is not dependent on the degree of malignancy. A more favorable response to therapy as regards the duration of the effect is usual in tumors of lower grades of malignancy. The less malignant the tumor, the longer the duration of life after the development of metastases. The course of the bone metastases in carcinoma is in general similar to the course of the primary neoplasm. In cases of latent or asymptomatic primary carcinoma manifested by metastatic lesions, roentgen study is of inestimable assistance in the search for the primary site. A thorough knowledge of the common and usual sites of metastases from the various types of malignancies is essential. Abrams analyzed 1000 consecutive cases of malignant neoplasms of epithelial origin autopsied between 1943 and 1947 at the Montefiore Hospital and found that 27.2 per cent had metastasized to bone. The skeletal metastases occurred in the following order of frequency:

	Per Cent of the Cases		*Per Cent of the Cases*
Prostate	84	Rectum	12.5
Breast	73	Stomach	11
Thyroid	50	Ovary	9
Lung	32.5	Colon	9
Bladder	26	Esophagus	12
Kidney	23.5	Tongue	9
Uterus	22	Larynx	7
Pancreas	15.5	Cervix	7.5

Metastatic lesions spread through the blood and lymph streams, the former being the more common route. Involvement below the elbow and knee is rare, although numerous exceptions occur. Lymph borne metastases are usually to the bone adjacent to the primary lesion. The most common examples of this are metastatic involvement of the bones of the pelvis, femurs and spine in carcinoma of the prostate and of the ribs, spine and skull in carcinoma of the breast. The bones most frequently involved in metastatic carcinoma are the ribs, skull, vertebrae, pelvis, femurs and humerus. Certain malignant diseases apparently have predilection for particular bones. Carcinoma of the thyroid is most apt to spread to the ribs. Hypernephroma spreads to the skull, ribs and long bones; the metastases are usually of the osteolytic type, single or multiple, often attain large size and may give clinical evidence of their presence before the primary tumor, many of which are asymptomatic for long periods of time. Ewing's tumor is the bone neoplasm which most commonly metastasizes to other bones. New bone formation may occur adjacent to the shaft of the bone and is parallel to the shaft or in the form of striations radiating from the bone and tends to be similar in appearance to the primary lesion. Bone lesions not infrequently result from direct extension of neoplasms in the adjacent tissue. Hypophyseal tumors derived from Rathke's pouch tend to invade the skeleton. Nasal polyps may

produce bone erosion by direct pressure. Carcinoma of the mouth and nose causes extensive destruction of the adjacent bone. Similar changes occur in carcinoma of the antrum. Lesions of the oropharynx may invade the base of the skull.

Primary sites such as the bladder, uterus, gastrointestinal tract and ovary are more apt to account for skeletal metastases than was previously thought to be the case. While it has been stated in the literature that gastrointestinal tract carcinoma metastasizes but rarely to bone, recent studies show that the contrary is true. The lesion in the gastrointestinal tract may be silent and give no manifestations of its presence. Careful study of the gastrointestinal tract with an opaque meal and barium enema must be carried out in all doubtful cases. It is not feasible to perform



Fig. 375. Metastatic Hypernephroma of the Femur. There is an extensive area of rarefaction in the upper third of the femur with a pathologic fracture.

roentgenographic studies of all the bones of the body in conjunction with barium studies and similarly at post mortem examination it is not practical to examine the entire skeleton in the absence of manifestations which indicate the possible presence of a bone lesion. The size of the lesion in the gastrointestinal tract appears to have no relation to the presence of metastases. It is not appreciated that bone metastases from cancer of the digestive tract are relatively common. The metastatic lesions in the bone may attain considerable size before producing pain or other symptoms. As a rule the lesions in the bone are purely osteolytic. The route by which distant metastasis frequently occurs is probably through the vertebral vein system described by Batson. Metastatic lesions of this type have been reported in the ribs, the skull, the long bones, and other parts of the body.



Fig 326 Metastatic Carcinoma of the Skull There is a rounded sharply defined area of increased radiance *in the superior parietal region* a typical metastatic lesion The primary lesion was in the breast



Fig 327



Fig 328

Fig 327 Metastatic Carcinoma of the Tenth Dorsal Vertebra *The body of the tenth dorsal vertebra is markedly* narrowed and irregular in outline There is also an area of rarefaction in the anterior aspect of the body of the ninth dorsal vertebra The primary tumor was a carcinoma of the breast

Fig 328 Metastatic Carcinoma of the Spine and Ribs Secondary to Carcinoma of the Stomach There is advanced metastatic disease *involving the* spine and the ribs The process is both osteoblastic and osteoclastic in character Several of the vertebræ *are partially* collapsed and show *marked narrowing* The primary lesion was in the stomach an adenocarcinoma



Fig 329 Metastatic Carcinoma from the Prostate There is advanced metastic carcinoma of the osteoclastic type with a pathologic fracture and intrapelvic protusion of the head of the femur The metastatic disease involves the ilium the pubis and the ischium on the left



Fig 330 Osteoblastic Metastatic Carcinoma There is increased density and irregular mottling involving the bones of the spine pelvis and hips The changes are characteristic of metastatic carcinoma of the osteoblastic type

In osteoclastic carcinoma, there are multiple areas of bone destruction scattered throughout the bones with no evidences of periosteal reaction expansion or cortical thickening Solitary lesions may attain very large size and destroy a great portion of the bone with pathologic fracture or collapse of the affected bone The osteoblastic lesions show areas of increased density throughout the involved areas The dense, spongy bone is the result of local osteogenesis The bone is increased in strength and pathologic fracture rarely results An extremely dense, uniform change in the bones may develop The picture may be closely similar to that in

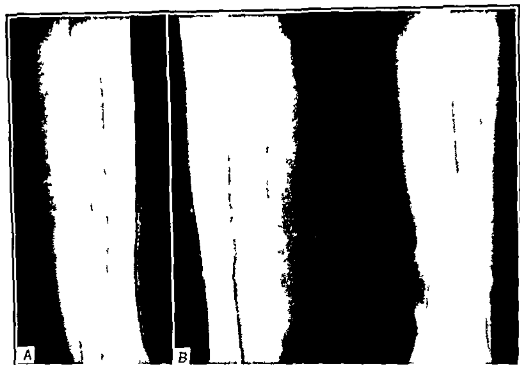


Fig 331 Osteoblastic Metastatic Carcinoma from Primary Carcinoma of the Bladder *A* There is a proliferative lesion in the lower third of the fibula *B* Eight months later the lesion has progressed and there are evidences of simultaneous bone destruction and new bone formation

Paget's disease The involvement may be diffuse with lesions in many bones and throughout the entire spine, in other instances only a single bone or one vertebral body is involved Carcinoma may produce osteoblastic lesions in the early stages and have extensive bone destruction in the later stages In some instances both bone production and destruction are present simultaneously Paget's disease may present a difficult problem in differential diagnosis as extensive sclerosis of bone may occur in osteitis deformans The characteristic feature of Paget's disease is subperiosteal deposition of osteoid tissue This widens the cortical portion of the bone with resultant enlargement of the bone shadow There is no enlargement of the bone in the presence of metastatic carcinoma and all evidences of bone trabeculæ are eliminated, whereas in Paget's disease, there is actual accentuation of the trabecular elements with a densely striated appearance Syphilis may cause osteosclerosis The bone is more homogeneously dense than in metastatic carcinoma

The Differentiation of Myeloid Metaplasia (Aleukemic Myelosis) and Metastatic Carcinoma of Bone

Myeloid metaplasia is a clinical and pathological syndrome characterized by weakness, hemorrhagic tendency, splenomegaly, hepatomegaly, aleukemoid peripheral blood with normoblastosis, and a specific type of myeloid metaplasia of the spleen and the liver. The condition is characterized by the constant occurrence of extramedullary hematopoiesis in the spleen and usually in the liver. There is an anemia with immature red and white cells in the peripheral blood. The condition very closely resembles leukemia and is frequently diagnosed as such. The diagnosis of myeloid metaplasia can be established with definiteness only at autopsy or by study of the marrow. The condition is known in the literature under various names, the most common being non-leukemic myelosis, myelosclerosis, aleukemic myelosis, and erythroleukemia. Myeloid metaplasia comprises non-leukemic extramedullary hematopoiesis. It is more common in the older age groups. Weakness is an outstanding symptom, most probably due to the anemia which accompanies the disease. The condition occurs in carcinoma and in many instances there are bone changes suggestive of the presence of osseous metastases. The combination of a primary carcinoma, the abnormal roentgen appearance of the bone, the erythroblasts in the peripheral blood, and the anemia are characteristic of myeloid metaplasia.

On histopathologic examination, the marrow is fibrous or shows areas of fibrosis and hyperplasia. Osteosclerosis is also present. Myeloid metaplasia is divided into two types: the primary without a known etiologic basis, and the secondary form. In the primary variety there is an exceedingly active marrow with predominance of normoblasts. Secondary myeloid metaplasia occurs in diseases of the bone marrow such as metastatic carcinoma, marble bone disease, myelofibrosis, osteosclerosis and tuberculosis. The osteosclerosis may be idiopathic or may represent a terminal stage of polycythemia rubra vera. The peripheral blood may have a high, low, or normal white cell count with large numbers of inter granulocytes, a condition identical to that which occurs in leukemia. The bone marrow may be fibrotic, hyperplastic or the site of carcinomatous metastases. The condition is believed to be a nonspecific response of immature multipotent cells of the liver and spleen to a wide variety of stimuli and is fundamentally different from leukemia.

There is aplasia of the bone marrow, replacement by fibrous tissue, and in the late stage the development of sclerotic changes. Enlargement of the spleen is probably a compensatory mechanism. The condition usually occurs in adult patients with no symptoms referable to the skeletal system. The blood picture simulates aleukemic leukemia. After the condition has been present for a period of years, roentgen studies show progressive sclerotic involvement of the entire skeletal system. There is no broadening of the shafts, coarsening of the trabeculæ, or visible periosteal reaction. The differential diagnosis must include primary osteopetrosis of Albers-Schonberg, osteosclerotic changes associated with leukemia, polycythemia, lymphogranulomatosis and Gaucher's disease. Also it is necessary to include the osteosclerotic lesions which follow alkaptonuria, fluorine, lead, phosphorus and arsenic poisoning and metastatic carcinoma of bone. In aleukemic leukemia with immature forms of both red and

white cells and associated anemia, roentgen studies may show alterations in the bones. In the early stages there is increased density of the ribs and other bones. This is followed by sclerosis, involving most of the bones. There is obliteration of the medullary space. Widening of the shafts, spontaneous fracture and periosteal reaction do not occur. Differentiation of osteoblastic metastatic carcinoma and myelosclerosis is not possible solely on the basis of the roentgen studies.

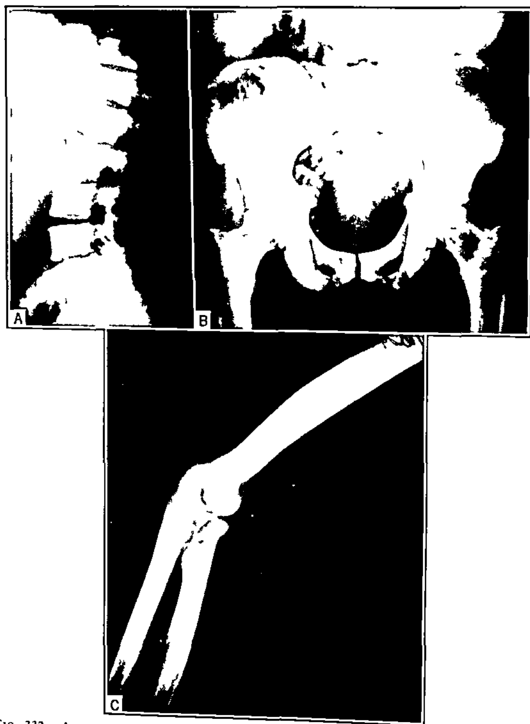


Fig 332. Agnogenic Myeloid Metaplasia. *A*, Lateral view of the spine. *B*, The pelvis. *C*, The right elbow. The bones show markedly increased density with coarse irregular trabeculations. The changes are generalized but are most pronounced in the spine and the pelvis. The patient had splenomegaly and evidence of extramedullary hematopoiesis. There was a moderate anemia with slight elevation of the white blood cell count.

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An understanding of the physiologic mechanism of this plexus makes clear the persistence into adult form of the embryonic type of vessels which exist in this region. All veins, particularly those which form a part of a large vascular bed, are low pressure systems and the flow of blood in this network is not necessarily in a constant direction. The flow may be to or from the cavities and it may be up or down the spine, depending upon muscle action, the force of gravity and other factors. Once carcinomatous material has gained entrance into the lumens of these vessels, the material can be transmitted to produce metastases in the vertebral column, the ribs, the skull, and other parts of the body without the lungs being affected.

Prior to the explanation by Batson, various theories were necessary to explain the seemingly paradoxical metastases. It was suggested that the foramen ovale remained open allowing metastases to enter by way of the venous system, transfer to the left side of the heart and be distributed by the arteries without involvement of the lung fields. The existence of differential filters has been suggested. This would necessitate the lung having a capillary bed of such a character that metastatic cells could pass through to be arrested in the capillaries of other tissues. It has also been postulated that there is a peculiar susceptibility of certain tissues and a predilection of the cells for such tissue. It has been found that as high as 40 per cent of metastases are paradoxical. Therefore, Batson's concept of a vertebral vein system which parallels the pulmonary and caval systems offers the logical answer. The presence of metastases to the lungs does not indicate that they have necessarily arrived there directly from the primary growth. The metastases may reach the lung from a metastatic lesion already established elsewhere in the body.

Batson endeavored to study the type of spread possible from the female breast by injecting small venules in the vicinity of the areolar mammae. He found in this experiment that there were two types of entirely unexpected distribution. It was demonstrated that the capillary venous plexus of the skin would carry more material than had previously been supposed possible. He was able to inject 30 cc. of roentgen opaque medium into the breast before the material appeared in the large veins. It spread in the subpapillary venous plexus and also to the opposite breast. The injected material entered the larger veins and flowed to the vessels of the axillary fossa, the veins of the clavicle, the vertebrae and the head of the humerus, and the larger venous sinuses of the skull. It also spread along the vasa vasorum of the upper extremities toward the elbow. On injection of the prostatic plexus, the material passed along the vasa vasorum to the level of the knee. This indicates that absence of metastases distal to the knee or elbow is due to temperature gradients or a failure of the material to be propelled along the veins distal to this point.

There are only a few isolated instances in which a metastasis has actually been found while in transit. Meyer Wildessen isolated a "string like mass floating free in a vein in a lower quadrant of the abdomen while removing a primary tumor. Henz reports a similar experience with a metastasis leaving the thyroid gland. The metastatic lesion in both instances was in a vein and the site of the primary neoplasm was definitely known. As a rule the site and nature of a primary neoplasm can be established. In the case of metastatic foci, however, only those in the regional lymph nodes can be identified with any degree of certainty. Lesions beyond these areas may be assumed to have originated and spread in a certain manner, but actual proof of the route cannot be established.

The Vertebral System as a Mechanism for the Spread of Metastases

The irregular distribution of disease has always been a matter of speculation and interest to clinicians The spread of malignant tumors is a field of the utmost importance In the case of metastases of tumors and abscesses, material is transported from the primary site to other parts of the body, either near or remote It has been proven that cells are the cause of this spread In consequence, the cells must move from one spot to another It is believed that the movement is through the blood or the lymph vascular system The concept of the blood vascular system consisting of a greater or systemic circulation and a lesser or pulmonary circulation with a lymphatic system joining the venous side has failed to explain the peculiar pattern found in the spread of malignant disease and pyogenic abscess There has been much speculation about the peculiarity of metastases of primary carcinoma of the prostate and the seminal vesicles In these diseases there is extensive invasion of the pelvis and vertebral column The pattern of the spread does not correspond to the lymphatic drainage of this region The pattern of spread of prostatic carcinoma mirrors the pattern of the extensive plexus of veins in the pelvis and the soft tissues about it and the plexus of veins in and about the vertebral column This plexus of veins is for the most part without valves The vessels are of considerable caliber and are found in the *epidural fat tissue as well as in the muscles and bones of the vertebrae* themselves

To study this plexus of veins Batson made use of the anatomic fact that the deep dorsal vein of the penis is an integral part of the prostatic vein plexus Injection of this vein affords access to the entire pelvic plexus and material so injected can be followed in its course around the walls of the pelvis along the rich plexus of veins With a thick injection mass the material tended to flow up into the inferior vena cava while with a thin watery mass the materials spread along the vertebral vein plexus The plexus for the most part lies at a lower hydrostatic level than that of the inferior vena cava The injection indicated the possibility of the spread along these vessels but did not demonstrate what happens in the living Similar injections were made in live monkeys The simple injection of roentgen opaque material did not cause a filling of the vertebral vein plexus the material ascending into the inferior vena cava With a tight binder placed around the abdomen and the injections repeated, the material was blocked in its course along the inferior vena cava and entered the vertebral vein plexus In the living animal there was flow from the *prostatic vein plexus into the vertebral vein plexus affording a mechanism* to explain the pattern of spread of carcinoma of the prostate The vertebral vein plexus with its tributaries and extensions form the large venous lake or reservoir which is connected with the veins of the thoraco abdominal cavity at each vertebral level The thoraco abdominal cavity is drained by the inferior vena cava and the azygos vein systems These course within the cavity to enter the heart At every body segment there are communications with the vertebral vein plexus The latter is protected in the bodies of the vertebrae in the deep back muscles and in the spinal canal and is not subjected to pressure changes of the body cavity as is the case with the vena cava and the azygos vein Therefore on compression of the thoraco abdominal wall the connections are able to bypass venous blood outside these cavities into the large plexiform venous bed

ness many being able to continue their regular occupations for considerable periods with consequent benefit not only to the individual but to the family and the community (6) there is striking betterment of the patient's mental status with helpfulness and optimism replacing the feeling of hopelessness and despair which so commonly occurs

The technique of roentgen sterilization varies in its details but in general follows fairly well established rules The object is to produce the desired result without injury to the skin and with as little disturbance of the patient as possible In our clinic the total dosage is usually 2 000 to 3,600 r varying with the size of the patient Two to four fields each measuring about 15 by 15 cm are used over the lower portions of the abdomen and lower back the rays being directed into the pelvis With larger dosages a permanent sterilization results and in most instances is followed by other symptoms which may accompany the menopause With smaller amounts of radiation the periods may return after an interval of six to eighteen months which is undesirable and may necessitate another course of treatment

Some authors have favored routine ovarian sterilization of all patients with cancer of the breast in the premenopausal age periods In the light of present day knowledge, this is not advisable Ahlbom in a very detailed study, showed that no definite benefits either as regards prolongation of life or the lessening of the incidence of metastatic lesions occur after ovarian sterilization Moreover patients suffering from cancer of the breast particularly those who have been operated upon, should not have the added burden of a premature menopause unless it is reasonably certain that definite benefit is to be expected from the procedure It is our experience that while ovarian sterilization has been followed by regression or total disappearance of osseous metastases it has not prevented new metastatic lesions from appearing Metastases in the bones have disappeared after roentgen irradiation of the pelvis only at a subsequent date to have new areas of destruction reappear in different portions of the skeletal system

It is our usual procedure to observe all patients with cancer of the breast carefully at frequent intervals after operation or radiation therapy Roentgen search for metastatic foci is carried out routinely two or three times annually This usually comprises roentgenograms of the chest skull, spine pelvis and upper femurs Roentgen examinations of the other bones are also made if there is pain, swelling or other clinical indication At the first evidence of the development of bone metastases roentgen sterilization should be performed Roentgen therapy directed to the involved areas may be administered simultaneously in an attempt to speed the regression of the lesions, although in some instances the metastases regress or disappear without such local therapy The general consensus is that about one third or more of the cases treated may be expected to benefit from this form of treatment In numerous instances, the benefits after sterilization were so striking that, though only an occasional case responded favorably the treatment would nevertheless be very much worth while There are no contraindications to ovarian sterilization and it may be used whenever there is any reasonable hope of benefit Since pregnancy is contraindicated in patients with metastatic lesions this is an additional benefit incidental to sterilization Patients who are markedly cachectic should receive smaller daily doses and have the treatments distributed over a longer period than those in good physical condition

The theory that the spread has been to the lymph node, heart, lung and blood stream is not applicable in every instance as in many cases metastases are present in the skull and brain yet the lungs are not affected

The vertebral venous system is a rich complex of large, thin-walled veins which surround the dural sac, invade the substance of the vertebræ and connect at each body segment through large segmented veins with the caval and azygos systems The cranial dural sinuses and the cerebral veins comprise a continuation superiorly of this system The veins of the head and neck may also be included as they are in the majority of instances valveless, and this is also true of the great mass of body-wall veins This large valveless meshwork is not encompassed by the thoraco abdominal cavity, receiving blood when the caval, azygos portal and pulmonary systems are compressed The vertebral complex parallels and bypasses the venous systems and their several parts Actual demonstration of the *proof of the existence of this venous system has proven difficult Batson's* work establishes the vertebral veins as an active and important route in the flow of blood and affords a logical explanation of a phenomenon which has puzzled all workers in the field of malignancy

Regression of Bone Metastases from Breast Cancer after Ovarian Sterilization

An extensive literature has accumulated in recent years stressing the importance of ovarian sterilization in the treatment of metastatic lesions from cancer of the breast Local recurrences and deep, visceral metastases generally seem to show little response after radiation treatment directed to the ovaries although occasionally very striking benefits in cases of these types are recorded in the literature The most satisfactory results seem to occur in patients with osseous metastases The rationale of the method is well established and in properly selected cases there is definite value in this mode of therapy The absence or withdrawal of the ovarian hormone is apparently the important factor in producing the favorable results In many instances roentgen treatment directed to the pelvis with consequent sterilization has resulted in partial or complete recalcification of metastatic bone foci far removed from the area exposed to irradiation with no therapy directed to the local lesions The question as to whether this form of treatment results in appreciable prolongation of life is still in doubt and cannot at present be definitely answered in the affirmative There has been unquestionable prolongation of life in some instances while in other cases there has apparently been no appreciable improvement in this respect

There are however certain definite benefits which may be expected in properly selected cases The chief among these can be summarized briefly as follows (1) there is usually prompt and effective relief of pain this may be of considerable duration and frequently lessens or temporarily obviates the need for morphia and other habit forming narcotics, (2) the lesions in the bones regress partially and in some instances entirely re ossify to disappear completely (3) fractures with their resultant deformities and prolonged disabilities may be prevented (4) there is great improvement in the patient s general condition with increased appetite and gain in wieght, (5) there is marked prolongation of the patient's useful-

Pathological Fractures in Bone Tumors

Pathological fractures may complicate benign and malignant primary or metastatic neoplasm and many other bone diseases The fracture is reduced and immobilized and the underlying disease is treated according to its nature Fracture through a primary malignant tumor such as an osteogenic sarcoma is of grave prognostic import as it is indicative of the fact that the lesion is advanced and extensive In many instances amputation becomes necessary and the fracture requires little or no treatment However, in the case of radiosensitive neoplasms such as endothelioma reticulum cell sarcoma and myeloma, the fracture requires the same treatment as uncomplicated fractures The most common type of pathological fracture is that associated with a metastatic lesion in the bone If the primary tumor is in the breast or the prostate, the response to hormones and roentgen treatment justifies the use of irradiation Many pathological fractures heal well

Sarcoma Metastases

Osteogenic sarcoma is in most instances a solitary bone tumor While metastases to other bones occur in some forms of this neoplasm, they are very rare, only very few cases of multiple osteogenic sarcoma having been recorded In a case reported by White there were multiple pulsating bone tumors with involvement of the calcaneus, the femur and the tibia of one extremity, the lesions developing successively in the various areas Microscopic studies of the amputated limb revealed evidence of osteogenic sarcoma In a case recorded by Silverman there were multiple osteogenic sarcomas in a twenty seven-year old male, the lesions involving the bones of the skull, the sternum, the vertebræ, the sacrum and the iliac bones Instances have been reported of multiple tumors involving the calvaria, the right femur, the left clavicle the scapula, the ribs and the dorsal and lumbar vertebræ Multiple skeletal tumors are commonly seen in the myelomas, and may occur rarely in Ewing s tumor and giant cell tumors These do not represent skeletal metastasis from a single primary bone tumor There is no criterion to differentiate multiple primary osteogenic sarcomas from skeletal metastases originating from a single primary bone tumor Osteogenic potentiality is frequently retained by secondary deposits in internal organs and tumor thrombi invading blood vessels There is no evidence to indicate that skeletal metastases in osteogenic sarcoma differ from those in other neoplasms It is not possible to distinguish primary and secondary tumors on the basis of the histopathologic structure alone On roentgen study the bone lesions resemble each other so closely that clear differentiation is impossible Osteogenic sarcoma metastasizing through the blood stream is much more apt to produce pulmonary metastasis than distant bone lesions Ackerman suggests an important feature which may be of significance in determining whether multiple lesions represent metastases Bone lesions which develop as metastases of primary osteogenic sarcoma are apt to affect the metaphyseal ends of the shaft While lesions in this location do not necessarily represent osteogenic sarcoma metastates from other tumors usually involve the diaphysis of the long bones and the bones of the trunk The sympathicoblastoma or sympathetic neuroblastoma arises in the neural tissue of the adrenal medulla and tends to metastasize to the cranium orbital region long bones and spine The bone metastases are often symmetrical

CHANGES IN BONE TUMORS AFTER THERAPY

In benign giant cell tumor roentgen irradiation may result in extensive ossification of the lesion with decrease in the size of the lesion Bone sarcoma may show recalcification and shrinkage of the tumor after irradiation Ewing s tumor responds favorably with regression of the tumor for long periods of time Chondrosarcoma and osteogenic sarcoma also regress after irradiation In carcinomatous metastasis roentgen therapy in the osteolytic type not infrequently results in reossification Osteoblastic metastases may remain stationary or regress after local roentgen therapy Irradiation of the ovaries has produced very marked reossification of osteolytic metastases in carcinoma of the breast and castration has resulted in regression of bone lesions from carcinoma of the prostate (see pp 630 and 631)

Pathological Fractures in Bone Tumors

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The classification of malignant lymphoma has been established by Jackson and Parker, the following groups being recognized (1) reticulum cell sarcoma, (2) lymphosarcoma, (3) Hodgkin s granuloma, (4) Hodgkin's sarcoma, and (5) giant follicular lymphoma

Hodgkin's granuloma is the most frequent form of malignant lymphoma approximately one third of the cases being in this group Hodgkin's sarcoma and giant follicular lymphoma are the least common and comprise less than 10 per cent of the cases The incidence of bone involvement varies widely In a series of cases reported by Coles and Schulz, 13 per cent of the patients showed lesions in the bones The giant follicular



FIG 333 Lymphoblastoma There is extensive rarefaction of the upper portion of the shaft of the humerus with a pathologic fracture The fragments are widely separated and markedly displaced The bone changes occurred in association with lymphoblastoma involving the mediastinum and the glands in the axillas neck and groin

lymphoma shows bone involvement only rarely Reticulum cell sarcoma which shows the highest incidence of bone lesions is discussed on p 618 Hodgkin s granuloma lymphosarcoma and Hodgkin s sarcoma are less apt to present osseous manifestations

The ages of the patients with bone lesions range from seven to eighty three years There is a slightly higher incidence of bone lesions in patients in the fourth to the sixth decades In many of the reported cases there is an apparent indication that younger patients are more apt to have lesions of bone This may be due to the greater life expectancy of this group The incidence in males and females is about equal It is rare for a patient with Hodgkin's disease to have attention first called to the disease by the bone lesion The symptoms referable to the skeletal system may develop at any time during the course of the disease The time of appearance of bone

involvement is indeterminate. In rare instances bone involvement occurs early and in these cases long term survival after adequate treatment is frequent. In Hodgkin's granuloma bone involvement tends to develop relatively late in the course of the disease. Autopsy studies indicate that there is a much higher incidence of bone involvement in Hodgkin's disease than appears to be the case as the result of roentgen studies. Therefore roentgen statistics are misleading in the study of this disease. This is due to the fact that the bone marrow may contain lesions which are too



Fig 334 Hodgkin's Disease A There is advanced destruction of the clavicle. *B* After roentgen therapy. The lesion is healed and there is extensive new bone formation



Fig 335

Fig 336

Fig 335 Malignant Lymphoma There is extensive destruction of the seventh cervical vertebra with collapse of the body of the vertebra. The disease was rapidly progressive. Postmortem studies showed malignant lymphoma with involvement of the mediastinal glands, lungs and bones

Fig 336 Lymphosarcoma of the Femur There is a pathologic fracture of the shaft of the femur in the region of the junction of the lower and middle thirds with marked bowing. There is extensive rarefaction of the bone in the region adjacent to the fracture. The patient was receiving deep roentgen therapy

small to involve the adjacent portions of the bone to a sufficient degree to be demonstrable by roentgen methods

Bone lesions in Hodgkin's disease are fairly common As a rule they are destructive, particularly in the skull and pelvis The punched out areas in these instances closely resemble multiple myeloma In the long bones the lesions are more common at the ends of the shafts In this instance, there is erosion of the cortex with expansion and periosteal changes Occasionally osteoblastic changes are present In many instances there is involvement of one bone, in others there are polyostotic lesions The bones commonly affected include the skull, spine pelvis sternum, ribs, extremities, and pectoral girdle The roentgen findings are very similar to those in metastatic carcinoma, osteogenic sarcoma bone cyst osteomyelitis, and multiple myeloma The bone lesions occur principally in the malignant type of the disease and are extremely resistant to irradiation therapy

It was formerly thought that Hodgkin s disease spread only through lymphatic tissue However, it is now known that the lesions of Hodgkin's disease may be spread through the reticulo endothelial system and some observers believe that the disease should be considered a disturbance of the reticulo endothelial system Skeletal involvement in Hodgkin s disease is indicative of marrow involvement The mechanism of the involvement of the bone marrow has not been demonstrated It is believed that it may take place by any one of three methods (1) direct invasion from adjoining granulomatous lesions, usually the lymph nodes, (2) hematogeneous invasion by means of small lymphogranulomatous emboli, and (3) transference of an agent capable of initiating a focus *in situ* This method could also initiate a primary lesion in the bone marrow It is essential to carry out roentgen and bone surveys on all patients with Hodgkin s disease Skeletal lesions are synonymous with marrow involvement the focus originating in the bone marrow and extending until it erodes the contiguous cortex of the bone In some instances the granuloma appears to invade the bone from adjacent lymphogranulomatous tissue This indicates usually that the disease is widespread and well advanced

No characteristic manifestations are produced by areas of bone involvement in Hodgkin s disease There may be osteolytic destruction or osteosclerosis In the case of the spine, there may be involvement of one or more vertebral bodies with collapse of the body Areas of increased density may be intermingled with areas of increased radiance Periosteal new bone formation is frequently present The osteoblastic reaction may be very marked In lymphosarcoma bone changes occur in about 10 per cent of the cases As in Hodgkin s disease the vertebral column is most frequently involved In many instances the patients complain of bone pain before localized osseous destruction or other changes are demonstrable by roentgen methods

In lymphosarcoma the long bones particularly the femur, the tibia and the humerus, are involved most commonly in individuals under forty The roentgen picture while not characteristic is of great assistance in establishing the diagnosis The condition is essentially osteolytic, patchy areas of bone destruction being intermingled with smaller areas of increased density There is no bone formation within the tumor However the changes in the periosteum may closely simulate those in osteogenic sarcoma when the tumor breaks through the cortex

Malignant Lymphoma of the Skull

Malignant lymphomas of the skull are rare. The lesion is usually discovered on roentgen examination made because of localized pain and swelling. It is a manifestation which occurs late in the course of the disease. Routine roentgenographic studies may disclose the existence of a bone lesion before there is any clinical evidence of its presence. The process is practically always osteoclastic in character, osteoblastic changes occurring only very rarely. Healing with recalcification may occur after roentgen therapy. Craver and Copeland in a review of 396 cases of malignant lymphoma found demonstrable changes in the bone in 15.7 per



FIG. 337. Malignant Lymphoma (Hodgkin's Disease). There is a large irregular area of increased radiance in the frontoparietal region. The margins of the lesion are markedly irregular and the advancing edges extend around small areas of normal bone which remain as sequestra or small islands of bone within the area of destruction.

cent of this group of patients. The bones involved in the order of their frequency were vertebræ, sternum, pelvis, femur, ribs, skull, humerus, scapula and clavicles. In the skull the lesions were in the parietal and frontal bones. In the case of the skull the roentgen picture is that of a single or multiple destructive lesions with irregular, poorly defined borders. The size of the lesion varies widely depending on the stage of the process at the time the bone involvement is discovered. Increased density may be present at the margins of the lesion although this is not the rule, the osteoclastic reaction usually predominating. There is no prominence of the blood vessel grooves and no evidence of increased intracranial pressure. The roentgen changes may closely simulate those due to many other conditions: Metastatic carcinoma, lymphosarcoma, leukemia, myeloma

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cases the vertebra assumes a biconcave shape termed the fish vertebra appearance. The x ray changes in the spine may precede the clinical manifestations by weeks or months and are of great importance in diagnosis.

Clinically, the diagnosis of leukemia in infants and children is more difficult than in adults since certain factors occur in children which do not appear of significance in adults. There is a tendency for non leukemic children to produce immature white blood cells in response to severe infections and frequently there is absence of leukocytosis in children with

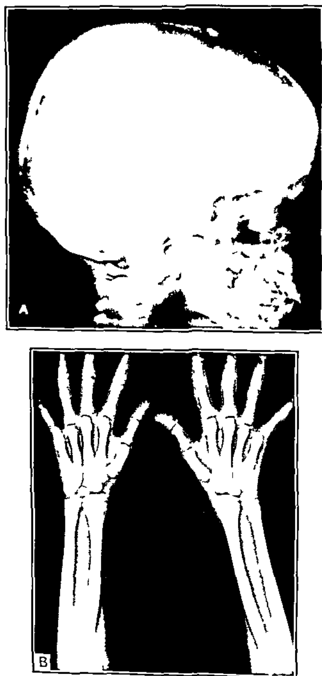


FIG 338 Leukemia *A* Lateral roentgenogram of the skull. The bones of the skull show diffuse granularity, generalized osteoporosis and thinning of the tables. *B* The bones of the fore arms, wrists and hands show cortical thinning, prominence of the trabeculæ and generalized increased radiance. The characteristic manifestations of a blood dyscrasia with hypertrophy of the marrow are well illustrated.

syphilis, osteomyelitis, eosinophilic granuloma, and fibrous dysplasia must be considered in the differential diagnosis The presence of Hodgkin's disease elsewhere in the body is an important factor in arriving at a diagnosis Biopsy may be required for final evaluation in doubtful cases

LEUKEMIA

Bone changes in leukemia are uncommon and may consist of bone destruction or bone production The former is more common Proliferative changes may occur in the periosteum or the endosteum In the case of involvement of the periosteum, the leukemic cells separate the periosteum from the bone The new bone formation particularly in the region of the skull, presents a pattern which has been described as resembling the teeth of a comb Endosteal hyperostosis is rare The bone changes are usually generalized The pattern is one of rapid alteration in appearance in accordance with the accelerated growth potential of the leukemic cells and the rapid remissions and exacerbations which are characteristic of the disease It is not possible by roentgen methods to differentiate between the lymphatic and myelogenous types Only the chronic form of the disease presents roentgen manifestations In children approximately half or more of the patients with chronic leukemia eventually show bone alteration Adults show advanced osteoporosis and focal bone destruction In advanced cases the leukemic osteoporosis may be very extreme with generalized loss of bone density There is associated elevation of the blood calcium in this phase of the disease Focal osteolysis may be scattered and widespread, the individual foci measuring only a few millimeters in size at the onset As the disease progresses, the lesions increase rapidly in size and the changes may closely simulate those in multiple myeloma Solitary lesions are rare In children, the manifestations are varied and comprise (1) juxta epiphyseal osteoporosis (2) generalized osteoporosis (3) scattered osteolysis (4) medullary sclerosis and (5) periosteal osteophytosis The changes in juxta epiphyseal osteoporosis are nonspecific and are characterized by a wavy band of increased radiance 1 0 mm or more in width extending across the immediate metaphyseal side of the epiphyseal plate due to tumor infiltration This does not develop until six weeks or more after the onset of the disease and may disappear within three weeks after the onset of a remission The involvement is most common in the proximal tibial, the distal femoral and the distal and proximal radial metaphyses The most common lesions are those of scattered osteolysis the so called moth eaten bones of leukemia The epiphyses and metaphyses of the long bones the small bones and the flat bones show round and oval defects These first involve the spongiosa and later destroy the compacta In advanced cases the picture is characteristic During remissions there is a condensation of the bone trabeculæ with osteosclerotic changes Periosteal new bone may be present in chronic leukemias of long duration and is manifested as a fine line of ossification parallel to the shaft of the diaphysis and separated from the original compacta by a clear space Medullary sclerosis is rare

Leukemic changes in the spine particularly in childhood, comprise productive and destructive lesions Bone destruction is manifested by a wedge shaped deformity and osteoporosis of the vertebral bodies This can be demonstrated relatively early by roentgen methods In advanced

different and the diagnosis can usually be established without difficulty Scurvy causes no confusion as other signs of scurvy are evident at the time the subperiosteal hematoma has formed Syphilitic periostitis is excluded by serologic study Infantile cortical hyperostosis and traumatic periostitis are differentiated by the clinical history and the blood picture Confusion with osteomyelitis is rare In Still s disease, there is thickening of the small tubular bones of the hands and feet which may simulate leukemia in its early stages The articular cartilages are not affected in leukemia while they are partially destroyed in Still s disease Osteoblastic metastases in carcinoma of the prostate may cause confusion in diagnosis The picture in Paget s disease differs from that in leukemia and study of the skull pelvis and other bones usually establishes the diagnosis Senile osteoporosis may produce changes which are similar to those in leukemia and differentiation by roentgen study alone is impossible Multiple myeloma similarly presents great difficulty in differential diagnosis

leukemia, hence the demonstration of leukemic changes in the bones by roentgen methods provides valuable information in establishing the diagnosis of leukemia during the early years of life There is a correlation between the roentgen appearances and the stage of the disease, the lesions being more numerous in the later stages The patients frequently complain of mild malaise or severe pain in one or several sites The pain may be in the bone and may be associated with local swelling The cause of pain is not known It is believed to be due to the pressure of proliferating leukemic cells on the adjacent nerves and blood vessels The rigid bone and tough periosteum do not permit expansion to the same degree as the soft tissue Intra osseous and subperiosteal pressure increase in proportion to the rapidity of proliferation The

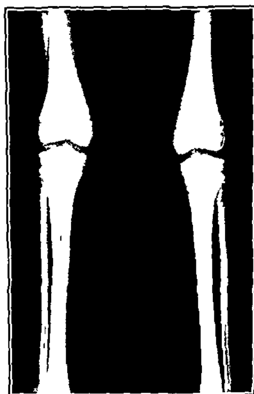


Fig 339 Leukemia Involving the Long Bones of a Child

transverse bands and destructive lesions are due to nutritional factors or focal bone destruction caused by pressure of the proliferating abnormal leukocytes The bony defects responsible for the transverse bands are reversible as the bands disappear spontaneously in many instances It has been shown by histopathologic examination that osteolytic changes are associated with the presence of large numbers of abnormal leukocytes in the bone The productive lesions, osteosclerosis and subperiosteal new bone formation are dependent upon stimulation of bone producing cells by the proliferating leukemic cells The osteoblasts of the periosteum form a layer of new bone over the masses of leukemic cells and separate the periosteum from its underlying cortex Stimulation of new bone formation results in osteosclerosis

Differential Diagnosis In the differential diagnosis metastatic neuroblastoma must be considered The clinical history of the two diseases is

cellular differentiation. The multiple foci of neuroblastoma which occur in the liver may be the result of multicentric origin of the neoplasm in cells in the liver which resemble neuroblasts rather than being due to metastases. The neuroblastoma has the same embryonic origin as the medulla of the suprarenal gland and the adjacent sympathetic ganglia arising from the neuroblasts which wander out from the neural crest during embryonic development to form the structures of the sympathetic nervous system. The primary growth may be small or attain very large size and tends to remain encapsulated. Early metastases are a prominent feature and all of the bones of the body may be involved. A severe anemia usually develops due to invasion of the bone marrow. The lesion is very highly malignant and terminates fatally despite surgery or radiation therapy.

Roentgen Manifestations. A. *Bone Lesions.* Practically all cases show medullary bone destruction, the margins of the involved area as a rule being poorly defined. Cortical destruction is often present and varies from a very small area to extensive erosion. The ribs, pelvis, vertebrae and long bones may be affected. The changes usually occur first along the medial aspect of the distal metaphysis of the femur or the proximal metaphysis of the humerus. Bilateral and symmetrical involvement is frequent, particularly in the diffuse fusiform lesions. Periosteal reactions occur in a high percentage of cases with elevation and thickening of the periosteum, usually parallel to the shaft, less commonly perpendicular or in sunburst formations. The bone lesions are oval, fusiform or diffuse. The metaphyses are involved less frequently than the diaphyseal portions. There is in many instances a soft tissue mass adjacent to the affected bone. Bone production usually occurs in association with destructive lesions. Generalized osteoporosis and epiphyseal changes are not present.

B. *Abdominal Changes.* An abdominal mass is present in many cases. Calcification may occur within the mass and may be punctate or extensive, solitary or multiple, homogeneous or mottled and dense or faint. The mass is frequently in the region of the adrenal and tends to displace the kidney downward and laterally. Enlargement of the liver is common.

C. *Chest Involvement.* The disease is frequently manifested by a rounded soft tissue mass in the posterior mediastinum. There is usually no pressure erosion of the adjacent bony structures such as occurs in neurogenic tumors of the paravertebral region. Metastatic lesions in the lungs may be in the form of nodules of varying size distributed irregularly throughout the lung fields. Pleural effusions, nodular thickenings of the pleura and masses in the mediastinum or the lung roots usually occur. Metastatic lesions in the spine and ribs are frequent. The lesions regress under roentgen therapy but tend to recur after a short interval of time. Calcification does not occur in the pulmonary or mediastinal lesions.

D. *Eye Tumors.* Soft tissue masses may occur in the orbit or facial region and may be associated with destructive lesions in the adjacent bones.

E. *The Skull.* In the skull there occur multiple small rounded areas of increased radiance due to bone destruction. These are most common in the parietal region. There is diffuse mottling, the lesions varying from very minute areas to a few millimeters in diameter with small confluent patches in some instances. The margins of the defects are slightly irregular and show no evidences of increased density. The sutures may be intact or widened, often to a considerable degree. The pituitary fossa is usually normal, although partial or complete destruction of the sella turcica has been

NEUROBLASTOMA

Peripheral neuroblastoma is a rare malignant neoplasm which usually arises from the sympathetic nervous system. The disease as a rule is rapidly fatal with extensive metastases to the bones, lymph nodes, liver, lungs, meninges and other organs. The condition may manifest itself as a tumor of the orbit or eye, the retinoblastomas being essentially similar to the neuroblastomas. It is frequently confused with Ewing's tumor, reticulum cell sarcoma and certain varieties of osteogenic sarcoma. Even histopathologic study may present great difficulties in establishing the diagnosis. The disease is most common in the period from the first few months of life to early childhood, only a few instances of congenital or

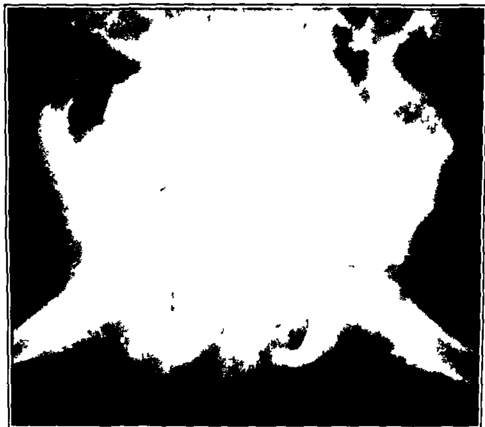


FIG 340 Neuroblastoma

adult lesions having been recorded. The average age is about four years. The duration of life in patients with neuroblastoma is about twenty one months. The most common sites of origin are within the abdomen, chest, neck, eye and orbit. In some instances the primary site cannot be determined even at autopsy. Bone lesions are common and are practically always multiple. An abdominal tumor occurs in the majority of cases. The thorax is among the most common sites.

Neuroblastomas develop as the result of abnormalities of differentiation of tissue and may arise in a single focus or simultaneously in multiple foci. Differentiation may continue with resultant alteration in the growth and character of the lesion. Thus a malignant neuroblastoma may mature into a benign form of ganglioneuroma as the result of continued

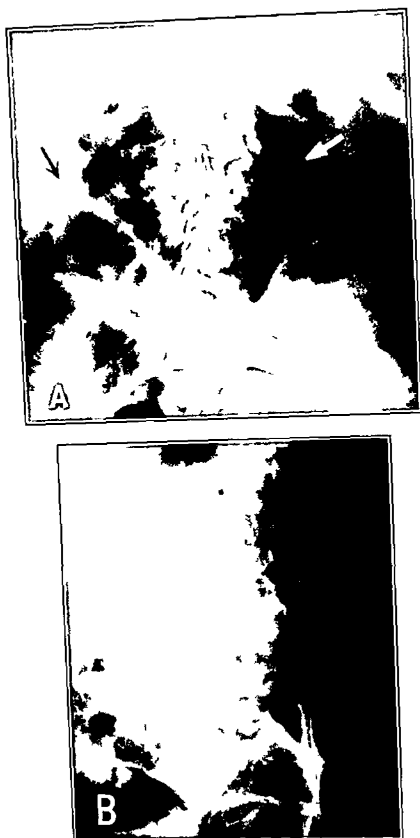


FIG 342 Chordoma Involving the Lumbar Spine *A* Anteroposterior view *B* Lateral view There is extensive destruction of the second third and fourth lumbar vertebræ with collapse of the vertebral bodies The distance between the twelfth rib and the crest of the ilium is markedly decreased because of the collapse of the lumbar vertebræ There is a large rounded soft tissue mass adjacent to the lumbar spine (arrows) The patient was a male sixty three years of age For fifteen years there had been pain in the back with gradually developing paralysis of the legs During the last two years of life there was a transverse myelitis with flaccid paralysis Postmortem diagnosis chordoma of lumbar spine with metastases to the lung

described Bone spicules may occur, extending outward from the decalcified outer table of the skull, particularly in association with soft tissue tumors of the scalp The base of the skull may be involved in the osteolytic process as well as the vault

Differential Diagnosis The characteristic roentgen manifestations comprise an abdominal mass, adenopathy orbital or eye tumor, and bilateral involvement of the bones in an infant or young child The skull manifestations are of great importance and frequently are sufficiently characteristic to establish the diagnosis The disease must be differentiated from Wilms' tumor, leukemia lymphosarcoma Hodgkin's disease, Ewing's tumor, osteogenic sarcoma hydronephrosis, osteomyelitis metastatic carcinoma, myeloma and the blood dyscrasias



Fig 341 Chordoma There is extensive destruction of the body of the fifth lumbar vertebra and upper sacrum on the left

CHORDOMAS

Chordomas are neoplasms which arise from remnants of the notochord It is a rare tumor and of relatively low malignancy When the lesion occurs in the region of the nasopharynx and the sacrum it may be detected by inspection and palpation In other portions of the body chordomas can be diagnosed as a rule by roentgen methods only prior to operation The lesions are being found with increasing frequency since the advent of the widespread use of the roentgen method of examination While chordomas may occur in any portion of the axial skeleton from within the skull to the coccyx they are most common at the extremities In a series of 252 cases collected from the literature by Faust and his associates approximately 37 per cent were cranial 13 per cent vertebral 48 per cent sacrococcygeal and the remaining few eccentric The tumors occur twice as commonly in men as in women The cranial chordomas appear most frequently during the third and fourth decades of life Those in the sacral region are most apt to occur in the fifth and sixth decades The fact that they are so rare in children has never been satisfactorily explained, particularly in view of the relationship to tissues which are prominent during fetal life

portions of the skull Very extensive destruction of the base of the skull is common The changes in the sella however, are usually slight or moderate in degree Calcifications occur only rarely Studies of the cervical spine may in the presence of a nasopharyngeal chordoma reveal a soft tissue mass with anterior displacement of the shadow of the air filled nasopharynx and areas of destruction in the bodies of the upper cervical vertebra

Displacement of the pineal shadow may be slight or marked Pneumoencephalograms and ventriculograms are helpful in demonstrating the tumor Tumors which arise in the region of the pituitary usually do not extend below the tentorium The position and contour of the intracranial portion of the tumor may be demonstrated by gas in the narrow subarachnoid cisterns The floor of the third ventricle is usually displaced upward while the aqueduct of Sylvius and the fourth ventricle are shifted

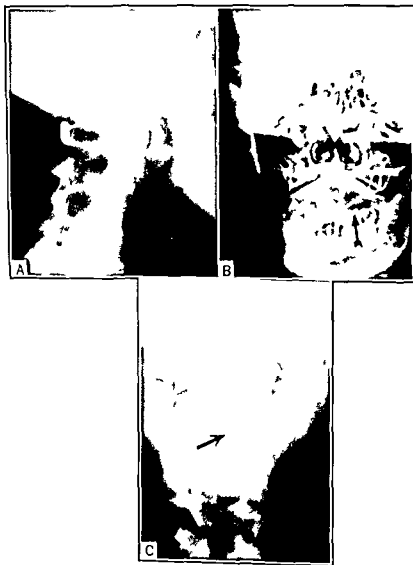


Fig 343 Chordoma with Extensive Destruction of the Cervical Vertebræ *A* Lateral projection Laminogram There is marked destruction of the posterior aspects of the bodies and pedicles of the second and third cervical vertebræ *B* Anteroposterior view The area of destruction involves the left side of the second and third cervical vertebræ *C* Anteroposterior laminogram The area of destruction is clearly defined The patient has a large soft tissue mass in the neck which projects into the oropharynx

Development The notochord forms the primitive central skeleton of all vertebrates and extends as a cylindrical rod from the buccopharyngeal membrane to the coccyx. As the embryo develops, mesodermal tissue surrounds the notochord to form the vertebræ and the intervertebral discs. The intravertebral segment disappears except for rare chordal vestiges. The intervertebral portion normally remains as the nucleus pulposus of the intervertebral discs. The cephalic part of the notochord forms a sigmoid curve through the basal plate of the skull in the mid sagittal plane. After regression cell rests may remain along the dorsal surface of the clivus intracranially, within the clivus, or in the retro pharyngeal tissues. Heterotopic chordal vestiges may undergo neoplastic metaplasia more readily than normal remnants as the topographic distribution corresponds closely to the sites of occurrence of chordomas. The ectopic vestiges of the notochord normally regress with advancing age. Chordal remnants occasionally are found during autopsy examination of the vertebral column. These may be placed posteriorly, centrally or less commonly laterally in the vertebral body.

Pathology The chordomas are slowly growing malignant tumors. In the typical case the neoplasm is rounded or lobulated and forms masses of soft gelatinous tissue with areas of hemorrhage, necrosis, cystic degeneration and calcification. At the onset the tumor tends to be encapsulated. As the lesion progresses the masses extend through the capsule and invade the adjacent bones and soft tissues. Fragments of detached cartilage and sequestrated bone are frequently present at the sites of invasion of bone by the tumor. On histopathologic examination the chordoma resembles the tissues of the embryonic notochord. The tumor is made up of large polyhedral cells with small nuclei and vacuolated cytoplasm which is distended with glycogen and mucin like material. The cells are arranged in cords, irregular columns or clusters separated by fine connective tissue trabeculæ. The most characteristic feature is the vacuolated appearance of the cytoplasm. Metastatic lesions have been *recorded only in tumors which arise in the region of the sacrum and* coccyx. The reason for this is not known.

The cephalic chordomas give rise to no characteristic syndrome. Intracranial protrusions of the growth may compress or distort the optic chiasm, other cranial nerves, the pituitary body, the crura, or even the pons. The general symptoms of elevated intracranial pressure may be late in appearing. Downward extension of the growth may provoke symptoms referable to the nose or nasopharynx. The neoplasm is soft and grows slowly so that in many instances it becomes very large before producing symptoms.

Roentgen Manifestations Cases have been reported in which the greater wing of the sphenoid has been destroyed, the defect extending to the ethmoid sinuses. The destruction may involve the foramen magnum or the lateral portion of the occipital bone. Lateral and sagittal projections of the skull may reveal destruction of the sella, particularly the dorsum sellæ and the posterior clinoids, the sphenoidal ridges and the walls of the sphenoidal sinuses. Areas of erosion in the basal and posterior portions of *the skull occur in the basiocciput and the petrous tip. The demonstration* of these changes requires carefully executed mentovertex, vertex mental and occipital (Towne) projections. The chordomas are primarily destructive of bone, and areas of increased radiance with complete absence of bony structure are demonstrable roentgenographically in the involved

ticularly in the region of the clivus. Cases have been reported with survival of nine or more years with external irradiation. The dosage must be high, in the neighborhood of 8,000 r or more. The treatment is given in series averaging 1,000 r each and repeated at intervals of a few months to a year. The ultimate prognosis of patients with chordoma is uniformly bad. Most cases survive for many months or several years, the average being three to four years.

PRE-SACRAL TUMORS

Pre-sacral tumors are rare. For purposes of description they are classified in four groups: (1) congenital anomalies, (2) bone tumors, (3) neurogenic tumors, and (4) miscellaneous. The most common congenital anomaly is the dermoid cyst. This lesion tends to remain silent until it has attained sufficient size to cause pressure. In most instances it does not manifest itself until adult life. The teratoma is a congenital anomaly which usually is present at birth. The mass is attached to the anterior surfaces of the sacrum or coccyx and grows forward and downward. Many neoplasms of infancy and childhood are congenital in origin. The growths develop as an abnormality of differentiation from groups of primordial cells which are pleuripotential or totipotential and are capable of producing two or three germ layers. The tumors are apt to occur at the sites of most active cellular differentiation. The sacrococcygeal tumor is one of the commonest of the neoplasms in the neonatal period. The neoplasm arises from the undifferentiated totipotential cells of the primitive knot, Hensen's node. As this node migrates toward the caudal extremity of the body, the cells which are later to form the genital cells are formed in the region of the urogenital ridge. Abnormalities of differentiation result in the development of teratomas anteriorly to the sacrum. Sacrococcygeal teratomatous tumors are most common in infancy and early childhood and are more frequent in females. The usual manifestations are a soft tissue mass in the region of the sacrum and coccyx. The tumor may reach enormous size compared to the patient's size and weight. The mass usually extends retrorectally. Calcification is frequent and when found in the presacral region aids in the differentiation from duplication of the colon and similar retrorectal masses. The calcific depostis are usually amorphous and irregularly scattered and do not indicate that the lesion is necessarily benign. Intravenous pyelography may demonstrate severe obstructive hydronephrosis and bilateral ureterectasis. The tumor may occupy the entire pelvis and extend into the abdomen. Metastases may be present in the lungs and skeleton. It must be differentiated from a meningocele. Meningocele communicates with the spinal cord and there is expansion of the mass when the patient coughs or cries, whereas teratomas show no such change. Anterior sacral meningocele is associated with anterior spina bifida. The sacro coccygeal chordoma is a progressive, slow growing tumor which invades and destroys bone by direct extension. The neoplasms of osseous origin comprise osteogenic sarcoma, osteochondroma, chondromyxosarcoma and giant cell tumor. The neurogenic tumors include ependymoma, neurofibroma, neuroclastoma and ganglioneuroma. The miscellaneous group consists of lipoma, fibroma, fibrosarcoma, sarcoma, hemangioma, lymphoblastoma, myeloma, metastatic neoplasms, ovarian cyst and abscess formations.

backwards The pons and medulla oblongata are displaced backwards and may be demonstrated between the gas shadows in the norrowed cisterna pontis and the fourth ventricle Obstruction to the circulation of the cerebral spinal fluid may result from compression of the mid brain at the incisura of the tentorium

The vertebral chordomas produce bone destruction due to infiltration by the tumor The vertebral bodies and arches are destroyed with eventual collapse of the vertebra The involvement of several adjacent vertebræ occurs more frequently with chordomas than with other types of malignant disease Myelography may disclose evidence of a tumor within the vertebral canal In this case there is obstruction to the flow of the opaque medium At the site of the obstruction, the subarachnoid space is narrowed in the gradual manner characteristic of extradural neoplasm

The roentgen manifestations of sacrococcygeal chordomas are typical There is extensive bone expansion which is best seen in the lateral projection This is a common and important feature in the early stages Destruction of bone may be pronounced at any time in the course of the disease A sharply defined soft tissue mass adjacent to or superimposed upon the sacrum may be demonstrable Within this mass a bony remnant or amorphous calcification may be present

The vertebral chordomas occur at any level and may extend into the soft tissues adjacent to the vertebral column Involvement of adjacent vertebræ and intervertebral discs is of diagnostic significance The lesion may easily be misinterpreted as being due to tuberculosis or pyogenic osteomyelitis, particularly in the presence of marked narrowing of the intervertebral disc A rare manifestation is localized osteoblastic change This may cause an erroneous diagnosis of metastatic carcinoma Hodgkin s disease, lymphosarcoma leukemia myeloma hemangioma, or osteitis deformans When the lesion is confined to one vertebral segment, as occasionally happens during the early stages there may be marked bone expansion and the lesion is easily mistaken for a benign giant cell tumor *Myelography is important to demonstrate the limits of epidural* extension of the tumor The tumor is very apt to extend from the segment of origin to an adjoining vertebra by growth through the intervertebral disc The primary roentgen change resulting from vertebral invasion is bone destruction The final stage of the process is compression fracture Expansion of the vertebral body and its transverse process may be an important feature The tumor does not remain confined to the vertebral column but extends into the adjacent soft tissues With discontinuity of the bony cortex posteriorly a soft tissue mass may not be demonstrable Tumors which extend anteriorly and laterally tend to be associated with a soft tissue mass In the presence of marked osteolysis, a soft tissue mass adjacent to the area of bone destruction and calcification or fragmented bone the diagnosis of chordoma may be established with a high degree of certainty

Treatment and Prognosis The treatment of chordomas is unsatisfactory Complete surgical excision is usually impossible because the tumor is in an inaccessible location and in close relationship to vital structures Surgery results in palliation and prolongation of life even with subtotal removal of large growths by relief of spinal cord compression in the presence of vertebral tumors Radiotherapy produces a favorable response in many instances although the results are palliative In some instances the tumor appears to be unusually radiosensitive, par-

ment to segment and each area observed accurately and in detail The dorsal region is more difficult to examine because of the kyphosis which is frequently present in this region The opaque material tends to separate into globules or discrete masses and its rate of flow can be controlled only with difficulty if at all in many instances In attempting to visualize the extreme upper segments of the cervical spine the contrast substance may enter the cisterna magna and the basal cisterns This should be avoided whenever possible It is essential that each intervertebral space be observed carefully and spot films made at each level, both with the opaque material ascending and descending On completion of the observations and the exposure of the desired roentgenograms the opaque material is returned to the lumbar sac and removed

The significant findings on the basis of which a diagnosis of an intraspinal lesion may be made by myelography are obstruction, deviation in the direction of flow a filling defect, marked asymmetry of the caudal end of the subarachnoid space narrowing of the subarachnoid space and deformity asymmetry, or absence of the axillary pouches or nerve root sleeves Multiple protrusions of the intervertebral discs may exist and these are demonstrable only by myelography Tumor may be present when a protruded disc is believed to be the cause of the symptoms and in rare instances both a tumor and a protruded disc may co exist in the same patient The absence of roentgen changes does not rule out the presence of a herniated disc or other spinal pathology This is particularly true in individuals in whom the subarachnoid space measures less than 16 mm Complete obstruction is rare, usually being produced by large mid line protrusions an extensive tumor or a marked arachnoiditis None of the opaque material passe the site of the block and the lesion frequently cannot be differentiated by roentgen methods, tumor and disc protrusion producing identical findings in many instances By introducing the opaque medium above and below the site of the obstruction the extent of the lesion can be demonstrated Defects narrowing of the opaque column and delay in passage may be due to mid-line disc protrusion lateral protrusion neoplasm arachnoiditis and other lesions

Myelography can be performed only by a closely knit team consisting of the neurosurgeon radiologist, nurse technician, and assistant Each must understand the others problems and cooperate fully throughout the procedure An intelligent and cooperative patient is also helpful After the conclusion of the injection the needle is best withdrawn as defects may be produced by a needle left in position In some clinics full reliance is placed on the roentgenoscopic observations and spot films made at this time Roentgenograms in the sagittal oblique and lateral projections made stereoscopically provide much additional data of value and should be taken routinely

Value of the *Plain Roentgenogram* The plain roentgenogram may supply very important data and is recommended routinely prior to myelography Expanding lesions in the spinal canal may be manifested by the roentgen demonstration of widening of the interpedicular spaces destruction or rarefaction of the lamina and pedicles, and changes in the intervertebral foramen These are usually late manifestations and are found in less than 20 per cent of the cases Herniations of the intervertebral discs may be associated with scoliosis straightening or reversal of the normal curvature *either complete* or segmental calcification narrowing of the intervertebral spaces encroachment on the intervertebral

Chapter

9

Diseases of the Spinal Canal

MYELOGRAPHY

Introduction The establishment of the syndrome of ruptured intervertebral disc by Mixter and Barr in 1934 as a cause of pain in the back has resulted in numerous methods being devised to demonstrate the lesion roentgenographically The procedure most commonly used is myelography, accomplished by the injection into the spinal canal of a contrast medium such as air a substance dissolved in oil, or a preparation that is water soluble Myelography is dependable in the diagnosis of protrusions of the intervertebral discs tumors of the spinal canal, arachnoiditis, and other conditions It permits of accurate localization of the lesion and establishes its extent and nature Approximately 85 per cent of spinal cord tumors and practically all cases of herniation of the intervertebral discs may be diagnosed by myelographic studies employing opaque media Early and accurate diagnosis of obstructive lesions is of the utmost importance to prevent progressive damage to the cord, to aid the surgeon in the determination of the level of operation and in planning the surgical approach

Air and aqueous preparations such as are used in intravenous urography have only a limited field of application although Lindblom and others recommend a water soluble medium in that it affords good visualization of the nerve root sheaths and is absorbed spontaneously Most clinics now utilize air or aqueous contrast media containing iodine only under special circumstances Pantopaque is the opaque medium most commonly used The injection is made into the lumbar region or the cisterna magna, the former being preferable as a rule Three to five cubic centimeters is the minimum amount recommended The procedure is carried out under fluoroscopic control By the use of a tilting table the progress of the contrast medium along the canal is observed The studies show whether an obstruction exists its level and the localization size and character of defects Complete blockage is usually due to spinal cord tumors but may also be produced by other lesions particularly arachnoiditis and herniated disc

Incomplete obstructions and filling defects must be interpreted with great care as globules of the opaque substance may apparently be obstructed by the dentate ligament nerve roots or inflammatory processes which involve the meninges In these instances there is not a true filling defect and the oil may be distributed in small collections along numerous vertebral segments In order to establish a diagnosis the deformity must be constantly present on successive examinations It is essential that the spinal canal be completely filled The lumbar region is the most satisfactory for study as the opaque material can be moved slowly from seg

the opaque medium to completely fill the lumbar canal with the patient in the erect position This converts the lumbar subarachnoid space into an opaque cylinder Deformations of the cylinder become demonstrable in profile, affording a much more satisfactory means of demonstrating a lesion than is possible when the canal is incompletely filled with the opaque material The method entails the use of 6—12 cubic centimeters in the average patient, although as much as 24 cc may be utilized if necessary The need for fluoroscopy is minimized as the roentgenograms provide all the necessary diagnostic data Very small defects may be masked Since the defects due to protruded intervertebral discs are as a rule peripheral rather than intraluminal in location even small lesions are demonstrable with correct projections The accuracy and speed of the examination are increased

PROTRUSIONS OF THE LUMBAR INTERVERTEBRAL DISCS

Most cases of protrusion of the lumbar intervertebral discs occur between the ages of twenty and forty The symptoms may be aggravated by posture movement and abnormal stresses of the muscles, ligaments and joints of the lumbar spine The manifestations referable to the lower limbs are caused by compression and stretching of the nerves The protruded disc tissue may be a large mass which fills the spinal canal with compression of all the nerve roots or a small mass of disc tissue may enter the canal and be in close relation to the anterior and posterior spinal root in the region of the point of emergence from the dural theca and entrance into the intervertebral foramen Sudden displacement of a mass of disc tissue compresses the nerve roots and the manifestations of a lesion of the lower lumbar and sacral roots ensue immediately The common etiologic factors are injury, pregnancy, labor, and marked weakness after an illness or operation In many cases no causative factor can be determined The history of injury usually includes lifting digging, a fall landing on the feet or buttocks direct trauma to the back or severe twisting movements The condition is commonest in those who do heavy manual work

The clinical manifestations may be divided into three main groups spinal tension and neurological The spinal and tension manifestations supply important information concerning the size of the protrusion and its relationship to the nerves and the theca The neurological signs indicate the severity of the injury to the nerve fibers and are important in determining which extradural nerves and intervertebral discs are affected The plain roentgenogram should be made in every instance to exclude lesions such as intraspinal tumor, primary or secondary vertebral neoplasm or tuberculous spondylitis The study may indicate the presence and level of the protrusion by showing narrowing of the intervertebral space with sclerosis of the adjacent vertebral surface and in some instances lipping of the margins of the adjacent vertebrae The value of the myelographic examination lies in the localization of the lesion and the determination as to whether it is single or multiple

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foramen by soft tissue masses or osteophytes and localized arthritis These changes occur in a wide variety of conditions and are not pathognomonic In performing myelography, it is essential to review the plain roentgenograms of the spine to determine the presence of anatomical variations, osteoarthritic changes, spondylolisthesis neoplasms and other lesions which may cause confusion in diagnosis

Technique of Roentgenoscopy After insertion of the needle, it is important to determine whether the subarachnoid space has been entered This is done by having the surgeon inject 0 5 to 1 cc of the opaque medium and observing roentgenoscopically whether the opaque material moves freely in the canal Pantopaque injected extradurally assumes a streaked appearance and opaque material injected into the subdural space is fixed or moves very slightly with change in position of the patient After the completion of the injection, the roentgenoscopic table is tilted into almost the upright position to determine the caudal extent of the cul de sac The opaque column is then moved cephalad by tipping the table slowly toward the Trendelenburg position As the opaque material reaches the lumbosacral and lumbar regions, the movement of the table is stopped and each interspace between the fifth lumbar and first dorsal levels is studied in the sagittal projections with half of the opaque material above and the remainder below the interspace under observation All suspicious areas should also be examined in the oblique and true lateral positions It is essential that the right and left oblique projections be made with the same degree of obliquity A landmark, such as the first sacral segment or the twelfth dorsal vertebra, should be included on each spot film The needle, which some surgeons prefer to leave in place may be used as the landmark if its exact level can be determined accurately A marker (usually a lead letter indicating R or L) should be on every film If it is found that 3 cc of the opaque material is insufficient to outline a suspected lesion it is well to inject an additional increment of 3 or more cc

The spot films should be developed immediately and observed while wet before the pantopaque is removed At the end of the examination the pantopaque is pooled so that the point of the needle lies centrally with relation to the opaque material In some instances, the needle point cannot be well visualized in the posteroanterior view A slight degree of obliquity reveals its position and direction The table may be tilted in one direction or the other to facilitate the removal of the opaque material After the opaque material has been removed the needle is withdrawn and a final spot film is taken to demonstrate residual amounts of pantopaque It is advisable to make 14" × 17' roentgenograms in the anteroposterior and lateral projections After the cervical spine has been examined, further roentgenograms should be taken for purposes of record These comprise lateral views of the skull and of the cervical region

Full Column Technique in Lumbar Myelography In the technique formerly used in myelography particularly with iodized oil small quantities of the opaque medium were found preferable Pantopaque however, is a lighter and less viscous substance which has the advantages of being relatively easy to remove from the spinal canal and more rapidly absorbed than lipiodol The use of 3 cc of pantopaque may prove unsatisfactory With the patient prone the opaque medium does not suffice to produce an unbroken column and false defects' may occur Defects in the nerve root sleeves may not be demonstrable Malis Newman and Wolf recommend a full column technique utilizing sufficient amounts of

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FIG 344 Herniation of Intervertebral Disc Myelogram with Pantopaque *A* Anteroposterior view *B* Oblique projection There is an irregular defect in the opaque column on the left at the level of the fifth lumbar vertebra and the first sacral segment The left nerve root sleeve at the level of the fifth lumbar vertebra is elevated blunted and shortened Operation revealed herniation of the intervertebral disc at the lumbosacral junction



FIG 345 Pantopaque Myelogram Herniation of the Intervertebral Disc at the Lumbosacral Junction *A* Anteroposterior view There is marked narrowing and irregularity of the opaque column at the level of the lumbosacral junction The defect in the opaque column is bilateral *B* Lateral projection There is a rounded defect in the anterior aspect of the opaque column at the lumbosacral junction Operation revealed an extensive herniation of the intervertebral disc

placed defects cause the opaque medium to assume an L or T shape. Osteoarthritic changes about the margins of the vertebrae may produce similar manifestations. The size of the defect is not a definite index of the size of the protrusion. A small defect does not necessarily mean a small lesion as the protrusion may be largely under the nerve root after its exit from the dural sac. Abnormalities of the nerve root sleeve occur frequently in lateral herniation of the disc and are due to compression of the nerve root sheath. In some instances, there is absence of the nerve root sheath. In others the nerve root sheath on the affected side is short, blunt, narrowed, irregular in outline, or elevated so that it lies at a higher level than its fellow on the opposite side. Displacement of the nerve root sheath indicates a change in the angle of exit of the nerve root. Widening of the caliber of the nerve root is due to edema. Rarely, there is a large unilateral lesion which exerts pressure on the subarachnoid portion of the subjacent cervical nerve root with medialward displacement.

Multiple herniations are not uncommon and may offer great difficulties in diagnosis. The protrusions may be ipsilateral, contralateral or in the mid line. Rupture of the disc and a tumor may be present simultaneously in the same patient. Recurrent herniations may occur and patients who have not been relieved by operation should be re-examined. In some instances a second, third or fourth protrusion may be discovered. Extramedullary tumors are usually associated with defects in the vertebral bodies and pedicles while herniations practically never produce these manifestations. Arteriovenous angiomas are manifested by dilated and tortuous arteries and veins. The enlarged vessels produce a characteristic serpentine radiolucent band in the opaque column. Angiomatous malformations of the spinal cord are less common than similar vascular anomalies in the brain. They may cause manifestations similar to those which occur in neoplasms. The so called varices of the cord are angiomatous malformations and may be venous or arterial. Non neurological lesions which may closely simulate protruded discs are arthritis of the spine, pelvic neoplasm, tuberculosis of the sacro iliac joints, osteoarthritic neuritis and herniated fat pads. In a series of cases reported by Young, symptoms which closely simulated protrusions of the disc occurred in patients with osteoid osteoma, glomus tumor of the leg, twisted ovarian cyst, multiple myeloma of the spinal column, eosinophilic granuloma of the pelvis, chondromyxosarcoma of the femur and tuberculous arthritis of the sacro iliac joint.

PROLAPSE OF THE DORSAL DISC

Statistical studies indicate that 5 per cent of all disc protrusions [illegible] multiple and involve more than one intervertebral space. The [illegible] combinations in the lumbar region are between the fourth and [illegible] vertebrae and between the fifth lumbar and first sacral [illegible] prolapse in the lower lumbar region and similar lesions in the [illegible] region at the level of the sixth and seventh cervical verte[illegible] frequent. The most common site of disc prolapse is in the [illegible] the lower cervical area being second in frequency [illegible] region being the least common. It has been estimated [illegible] dorsal region comprise approximately 5 per cent [illegible] complete examinations of the dorsal region more [illegible] found. In many of the cases, the prolapse of [illegible]

ciated with another prolapse The most frequent site is between the eleventh and twelfth dorsal vertebræ In many instances the lesion involves the twelfth dorsal and first lumbar segments Thoracic lesions above the level of the fifth thoracic vertebra are extremely uncommon

CERVICAL DISC PROTRUSIONS

Protrusion of the cervical discs is more common in the fourth to sixth decades of life and occurs more frequently in men The usual sites are between the fifth sixth and seventh cervical vertebra The distribution and character of the pain are bizarre Pain is frequently absent and when present is not severe There is weakness, paresthesia and numbness in both upper limbs The pain may be bilateral or localized to one arm Sensory disturbances may also be present The tendon reflexes are normal or slightly increased The symptoms are commonly referred to the dorsal region the pyramidal tracts and the lateral and ventral spinothalamic tracts Root pain is not a predominant symptom Partial or complete block may occur with elevation of the protein level in the cerebrospinal fluid Plain roentgenograms usually do not aid in the diagnosis The technique of the examination in the cervical region is more difficult than in other portions of the spine It is inadvisable to permit the opaque medium to enter the basal cisterns and cranial cavity The pantopaque is injected into the lumbar subarachnoid space and the patient lies prone with the neck extended With the patient held securely in position, the table is tilted and the movement of the opaque column is observed It may be necessary to invert the patient to an angle of 70 to 85 degrees for the medium to enter the upper thoracic and lower cervical region The movement of the table must be carefully controlled and if the pantopaque passes too rapidly into the cervical canal, the table should be returned promptly to the horizontal position

In the lumbar spine the opaque medium usually maintains itself as a solid column *In the thoracic region, there is a tendency for the medium* to separate into droplets This can be obviated by returning the patient to the erect position It is usually not possible to make a completely satisfactory examination unless the pantopaque is maintained in a single column Normally there is transitory hesitation at the region of the distal portion of the cervical canal After a short pause, the column as a rule passes along either side of the canal It outlines the axillary pouches as small triangular shadows with the apices directed laterally in the region of the cervical nerves as they pass through the intervertebral foramens It is difficult to fill the entire canal

Herniations of the nucleus pulposus in the lower cervical region may produce a filling defect In some instances the opaque medium halts briefly at the level of the lesion and forms a large globule with a flat or convex base directed cephalad A thin trickle appears at one or both sides of the lesion The appearance at the level of the lesion may be that of an inverted U or L with thread like droplets beneath and between the arms A ruptured intervertebral disc protruding into the lower cervical canal may cause no obstruction partial obstruction or complete block The picture may be very similar to that in spinal cord tumors with a cap like defect proximal to which lateral streaks extend for several segments along one or both sides of the canal Small triangular protrusions may be visualized between the pedicles due to the accumulation of opaque material

in the axillary pouches In the presence of a herniated disc in the lower cervical spine the cap like defect is transitory and may be overlooked easily The lateral streaks are shorter and the opaque material does not accumulate in the axillary pouches In the presence of complete blockage, the lesion cannot be distinguished from extramedullary tumor



FIG 346 Cervical Myelogram with Pantopaque Herniated Disc There is an irregular defect on the left at the level of the junction of the seventh cervical and first dorsal vertebræ The nerve root sleeve on this side is absent Operation revealed a herniated disc



FIG 347 Hypertrophy of the Ligamentum Flavum A Anteroposterior view There is a defect in the opaque column on the left side at the level of the fourth and fifth lumbar vertebræ *B* Lateral view The defect in the opaque column at the level of the fourth and fifth lumbar vertebræ lies posteriorly indicating that it is due to hypertrophy of the ligamentum flavum rather than to protrusion of the disc

ciated with another prolapse The most frequent site is between the eleventh and twelfth dorsal vertebræ In many instances the lesion involves the twelfth dorsal and first lumbar segments Thoracic lesions above the level of the fifth thoracic vertebra are extremely uncommon

CERVICAL DISC PROTRUSIONS

Protrusion of the cervical discs is more common in the fourth to sixth decades of life and occurs more frequently in men The usual sites are between the fifth sixth and seventh cervical vertebræ The distribution and character of the pain are bizarre Pain is frequently absent and when present is not severe There is weakness, paresthesia, and numbness in both upper limbs The pain may be bilateral or localized to one arm Sensory disturbances may also be present The tendon reflexes are normal or slightly increased The symptoms are commonly referred to the dorsal region the pyramidal tracts and the lateral and ventral spinothalamic tracts Root pain is not a predominant symptom Partial or complete block may occur with elevation of the protein level in the cerebrospinal fluid Plain roentgenograms usually do not aid in the diagnosis The technique of the examination in the cervical region is more difficult than in other portions of the spine It is inadvisable to permit the opaque medium to enter the basal cisterns and cranial cavity The pantopaque is injected into the lumbar subarachnoid space and the patient lies prone with the neck extended With the patient held securely in position, the table is tilted and the movement of the opaque column is observed It may be necessary to invert the patient to an angle of 70 to 85 degrees for the medium to enter the upper thoracic and lower cervical region The movement of the table must be carefully controlled and if the pantopaque passes too rapidly into the cervical canal the table should be returned promptly to the horizontal position

In the lumbar spine the opaque medium usually maintains itself as a solid column In the thoracic region there is a tendency for the medium to separate into droplets This can be obviated by returning the patient to the erect position It is usually not possible to make a completely satisfactory examination unless the pantopaque is maintained in a single column Normally there is transitory hesitation at the region of the distal portion of the cervical canal After a short pause, the column as a rule passes along either side of the canal It outlines the axillary pouches as small triangular shadows with the apices directed laterally in the region of the cervical nerves as they pass through the intervertebral foramens It is difficult to fill the entire canal

Herniations of the nucleus pulposus in the lower cervical region may produce a filling defect In some instances the opaque medium halts briefly at the level of the lesion and forms a large globule with a flat or convex base directed cephalad A thin trickle appears at one or both sides of the lesion The appearance at the level of the lesion may be that of an inverted L or L with thread like droplets beneath and between the arms A ruptured intervertebral disc protruding into the lower cervical canal may cause no obstruction partial obstruction or complete block The picture may be very similar to that in spinal cord tumors with a cap like defect proximal to which lateral streaks extend for several segments along one or both sides of the canal Small triangular protrusions may be visualized between the pedicles due to the accumulation of opaque material

in the axillary pouches In the presence of a herniated disc in the lower cervical spine the cap like defect is transitory and may be overlooked easily The lateral streaks are shorter and the opaque material does not accumulate in the axillary pouches In the presence of complete blockage, the lesion cannot be distinguished from extramedullary tumor



Fig 346 Cervical Myelogram with Pantopaque Herniated Disc There is an irregular defect on the left at the level of the junction of the seventh cervical and first dorsal vertebræ The nerve root sleeve on this side is absent Operation revealed *a herniated disc*



Fig 347 Hypertrophy of the Ligamentum Flavum A Anteroposterior view There is a defect in the opaque column on the left side at the level of the fourth and fifth lumbar vertebræ B Lateral view The defect in the opaque column at the level of the fourth and fifth lumbar vertebræ lies posteriorly indicating that it is due to hypertrophy of the ligamentum flavum rather than to protrusion of the disc

HYPERTROPHY OF THE LIGAMENTUM FLAVUM

Hypertrophy of the ligamentum flavum is more frequent in the male than in the female The lesion may occur at any age, although it is most common in the third and fourth decades of life There is usually a history of trauma The initial symptoms are pain in the back unaccompanied by nerve or superficial tenderness Bending, coughing, sneezing or lifting aggravate the pain The lesion is diagnosed by the use of the myelogram The defect produced by hypertrophy of the ligamentum flavum is similar to that in protrusion of the disc except that the changes are at the posterior aspect of the opaque column rather than laterally

ARACHNOIDITIS

Arachnoiditis is a proliferation of the leptomeninges which causes adhesions between the pia and arachnoid and cyst formations The etiology of the condition is not clearly understood Meningitis and trauma are the common causes The neurological symptoms which develop as a result of the associated changes in the nervous system are supposedly caused by mechanical injury secondary to the cysts and adhesions and do not ensue from the primary process which produced the cysts or adhesions Partial paraplegia, pain and loss of sensibility occur over the lower part of the body The level of the lesion is indicated by radicular pains paresthesias, and loss



Fig 348



Fig 349

Fig 348 Arachnoiditis with Obstruction at the Level of the Eleventh Dorsal Vertebra The opaque material was introduced in the lumbar region In the Trendelenburg position the opaque column meets with obstruction at the level of the eleventh dorsal vertebra There is marked irregularity of outline narrowing and mottling of the opaque column at the level of the obstruction Operation revealed arachnoiditis

Fig 349 Arachnoiditis Myelographic study shows irregular streaking of the opaque material with division of the column into numerous strands There is partial obstruction at the level of the inferior margin of the fifth lumbar vertebra

of sensation, although the upper limit of the anesthesia is seldom sharply defined as in cord tumor. It may be impossible to withdraw spinal fluid because of obliteration of the lumbar subarachnoid space. If fluid is obtained it shows elevation of protein but no increase in cells. Manometric studies reveal partial or complete obstruction of the spinal canal. Operation discloses a thickened pia-arachnoid which invests the cord closely and constricts the spinal nerve roots. The subarachnoid space is obliterated in part or completely. Small cysts are present in the meninges. The process may be localized or extended over many segments of the cord.

On myelographic study, the opaque medium may be arrested at the level of the lesion and the border of the obstruction is serrated and irregular, rather than smooth or rounded as in tumor. If the obstruction is incomplete, there are filling defects and multiple irregular, rounded or linear streaks of the opaque material in the involved portion of the canal with partial arrest or marked delay in the passage of the contrast substance through this area. The opaque column may be broken up into numerous discrete globules of varying size. Several segments of the spine are usually involved. Attempts to withdraw the injected opaque material after completion of the myelographic studies are usually not successful, small amounts being retained in the pockets formed by the adhesions and cysts.

SPINAL EXTRADURAL CYST OR DIVERTICULUM OF THE SPINAL ARACHNOID

The previously reported cases of extradural cysts comprised dermoids, epidermoids or lesions due to the echinococcus. Elsberg, Dyke, and Brewer in 1934 reported 4 cases of spinal extradural cyst which produced compression of the spinal cord and considered the condition a definite clinical entity. The cysts practically always occur during adolescence and are associated with kyphosis and epiphysitis of the thoracic portion of the spinal column. Most of the cases are in males under the age of twenty years. Symptoms may be present for months or years. The lesions are practically always located in the middle or lower thoracic region, although involvement of the cervical and lumbar areas has also been reported. The cysts originate laterally near the emergence of the spinal nerve roots. The manifestations are localized to the ipsilateral side, the other extremity being affected later. The condition clinically simulates multiple sclerosis or similar degenerative diseases.

Spinal extradural cysts may be of two types. Those which occur in adolescence involve the dorsal spine and are associated with kyphosis juvenilis while those which occur in adults are not accompanied by kyphosis. The origin of spinal extradural cysts has not been established. It is believed that the cyst is the result of a congenital diverticulum of the dura mater or a herniation of the arachnoid due to a congenital defect in the dura. There is a tendency for the lumbar extradural cysts to manifest themselves at a later age than those in the dorsal region. This is most probably due to the fact that the lumbar canal is spacious, permitting the cauda equina to be displaced so that the cyst can increase in size without producing symptoms for a longer period than in the dorsal area. It has been suggested that dorsal kyphosis juvenilis may be the result of venous congestion and stasis in the vertebral bodies. In many instances the clinical diagnosis is that of a herniated disc.

Roentgen Manifestations Roentgen study reveals changes which make definite diagnosis possible During childhood, there is enlargement of the spinal canal with the characteristic changes of kyphosis dorsalis juvenilis in the bodies of the involved vertebræ These findings apparently do not occur in any other condition In cases which develop after the bony development of the spine has been completed, kyphosis dorsalis juvenilis is not present The pedicles of more than one vertebra show erosion along their inner margins the changes extending the entire length of the cyst The vertebral bodies are wedged and irregular in outline anteriorly The erosion of the pedicles of the involved vertebræ is best demonstrated by the increased width of the interpedicular spaces There is usually flattening and narrowing of the medial aspects of the pedicles and enlargement of the intervertebral foramens These manifestations indicate the presence of a large mass within the spinal canal Myelography shows enlargement of the neural canal In some instances the opaque medium extends beyond the confines of the spinal theca and through the intervertebral foramens The passage of globules of the opaque material beyond the lateral limits of the neural canal indicates the presence of recesses or diverticulums which communicate with the subarachoid spaces In some instances there may be a communication between the lesion and the subarachnoid space the opaque material entering the cyst Extensive spina bifida, meningocele, or myelocele may be present There may be an associated diastematomyelia and a bony, cartilaginous, or fibrous process may protrude from the dorsal surface of one or more of the vertebral bodies in the form of a narrow band and partially or completely divide the spinal canal into two parts In these instances, myelographic studies show partial obstruction and a thin band extending through the shadow of the canal due to the fibrous or cartilaginous adhesion A bony defect or protrusion is best demonstrated by myelography

VASCULAR ANOMALIES OF THE SPINAL CORD INTRASPINAL HEMANGIOMAS AND DILATATIONS OF THE SPINAL CORD VEINS

Vascular anomalies of the central nervous system include both arterial and venous malformations Although most frequent in the brain they also occur in the spinal cord and the retina Bleeding may ensue and hemorrhage into the subarachnoid space may produce an acute transverse myelopathy In the previously reported cases there have been no associated cutaneous nevi which is in striking contrast with the reports of skin hemangiomas in venous or arteriovenous anomalies of the brain The neurological manifestations are characterized by disturbances of the sensory levels which shift during the same and on subsequent days There is evidence of progressive cord damage Any portion of the spine may be affected Venous dilatations of the spinal cord are difficult to diagnose Intraspinal vascular anomalies may occur in the form of dilatations of the pial veins as arterial or arteriovenous aneurysms or as hemangiomas Arterial and arteriovenous aneurysms cannot be distinguished from true venous dilatations Venous dilatations other than those associated with neoplasms are due to interference with the venous circulation of the spine with resultant varix formation Congenital anomalies of the development of the veins result in the production of angiomas

The dilated veins lie in the spinal canal and are surrounded by cerebrospinal fluid. Engorgement of the veins within the cord impedes the circulation of the spinal fluid. Since the veins may at one time be dilated and at others collapsed, a constant roentgen picture is not produced. The plain roentgenograms of the spine reveal no abnormalities as a rule, although rarely there may be erosion of the pedicles, the laminae, and the articular processes. The pedicles may become flattened or concave and the interpediculate measurements increased. The involvement as a rule is localized to one vertebral segment. Congenital anomalies of the vertebrae frequently are found in association with varices. On myelography, the varices produce a pattern of negative serpentine areas due to displacement of the opaque medium by the dilated veins. There is a mottled distribution of droplets of the opaque medium extending over one or more vertebral segments. The negative shadows within the spinal canal present smooth, parallel margins. In pial medullary angioma there is a constant filling defect characterized by multiple rounded and linear striations dis-

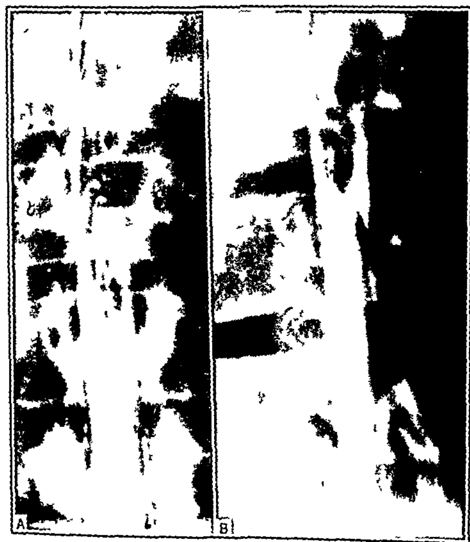


Fig 350 Varices in the Lower Dorsal and Upper Lumbar Region. Myelogram. *A* Anteroposterior roentgenogram. *B* Lateral projection. Within the opaque column there are multiple rounded and linear areas of increased radiance. On roentgenoscopic study the opaque material moved slowly and in a tortuous fashion in this region. At operation multiple varices were found within the spinal canal.

tributed irregularly within the opaque column which indicate that the opaque medium surrounds the vessels The presence of many sharply defined, worm like filling defects is characteristic and establishes the diagnosis Partial blockage may occur In adhesive arachnoiditis irregular deformities due to the presence of pseudocysts and cicatrices may present a closely similar picture Varicosities of the spinal canal veins may be intradural or pial and produce a roentgen picture very similar to that in disc herniation If the defect is not at the level of a disc, there is no problem in diagnosis A defect which is solitary and at the level of a disc creates a very difficult problem Varicosities tend to change in size or shape during the examination while the defect produced by a protruded disc remains constant Multiple defects are more common with varicosities

THE ARNOLD-CHIARI MALFORMATION

The Arnold Chiari malformation of the hindbrain is a protrusion of the cerebellum and brain stem through the foramen magnum into the cervical spinal canal It is a congenital defect and in many instances is associated with spina bifida, myelomeningocele and hydrocephalus The extent of the anomaly varies widely It may occur in the absence of skeletal or central nervous system defects or in association with bony abnormalities of the craniovertebral junction such as platybasia and fusion of the cervical vertebrae The condition has been found at all stages of life from the neonatal period to adulthood While the diagnosis can frequently be established on the basis of the clinical manifestations, the condition in other instances is discovered only at operation or autopsy Arnold in 1894 recorded the case of a newborn infant who showed protrusion of a portion of the cerebellum through the foramen magnum over the dorsal aspect of the upper cervical spinal cord Chiari divided the anomaly into three general types The first consisted of elongation of the cerebellar tonsils and inferior lobes with extension into the upper cervical canal The fourth ventricle was not displaced This variety occurs in both children and adults and may be associated with internal hydrocephalus The second is characterized by displacement of the fourth ventricle caudally and represents a more marked deformity Internal hydrocephalus and spina bifida are associated with this form The third type is most advanced There is downward displacement of the entire hydrocephalic cerebellum into the cervical spine canal The pathogenesis is still indeterminate It is believed to be the result of an abnormal relationship of the hindbrain to its bony envelope An abnormality of fixation of the spinal cord and its meningeal coverings and roots by a spina bifida may prevent the spinal cord from ascending normally as the spinal column grows and the cauda equina develops Hydrocephalus meningocele and spina bifida may also occur The greater the defect and the more caudal its location the earlier the appearance of the deformity and its resultant complications

The clinical manifestations are variable and inconstant The condition is usually confused with cerebellar tumor Various paralyses, headaches, ataxia, cranial nerve palsies visual disturbances parasthesias and other symptoms have been recorded Studies of the cerebrospinal fluid may reveal partial or complete block with elevation of the protein content There may be motor or reflex changes in the extremities nystagmus, and

cerebellar signs Diplopia, papilledema, deafness, vomiting and dysfunction of the ninth to twelfth cranial nerves are less frequently present The cerebellum is grossly abnormal and there is no vermis The medulla oblongata lies largely caudad to the foramen magnum The cervical nerve roots may incline upward in their intrathecal course Many observers believe that the condition is an anomaly of the hindbrain rather than secondary to mechanical factors Therapy comprises surgical relief of obstruction of the cerebrospinal fluid pathways This is best accomplished by moderate suboccipital craniectomy and upper cervical laminectomy The dura is opened widely Freeing of adhesions and resection of the cerebellar tongues is not indicated



FIG 351 Arnold Chiari Malformation Pneumoencephalogram There is narrowing of the air column in the region of the foramen magnum and the upper cervical vertebræ (arrow) the characteristic manifestation of protrusion of the cerebellum into the cervical spinal canal

Roentgen Manifestations Roentgen examination of the cervical spine may be negative or reveal some of the abnormalities described above On myelographic study there is a filling defect in the opaque column in the region of the cisterna magna and upper cervical region The defect is bilateral and is characterized by an indentation due to the accumulation of the opaque medium in the cleft between the herniated cerebellar tonsils List describes an arrest in the passage of the opaque material in the upper cervical region and a filling defect which presents a concave upper margin The change has been described as being due to a mass within the upper cervical area with a block to opaque material injected below this region The obstruction is less complete than with a tumor of similar size and the flow into the vault is less impeded than that in the reverse direction The edge of the mass shows lobulations due to the gyri of the herniated cere-

tributed irregularly within the opaque column which indicate that the opaque medium surrounds the vessels The presence of many sharply defined, worm like filling defects is characteristic and establishes the diagnosis Partial blockage may occur In adhesive arachnoiditis irregular deformities due to the presence of pseudocysts and cicatrices may present a closely similar picture Varicosities of the spinal canal veins may be intradural or pial and produce a roentgen picture very similar to that in disc herniation If the defect is not at the level of a disc, there is no problem in diagnosis A defect which is solitary and at the level of a disc creates a very difficult problem Varicosities tend to change in size or shape during the examination while the defect produced by a protruded disc remains constant Multiple defects are more common with varicosities

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SPINAL CORD TUMORS

It is estimated that 75 to 85 per cent of spinal cord tumors are benign and potentially curable hence it is of the utmost importance to establish the diagnosis as early as possible and especially before irreversible changes have occurred in the spinal cord Many of the cases are not diagnosed until late principally because the lesion had not been considered in differential diagnosis Tumors of the spinal cord are uncommon Careful attention to the clinical and roentgen manifestations makes definite diagnosis possible in approximately 95 per cent of the cases



Fig 352



Fig 353

Fig 352 Neurofibroma Myelography with pantopaque shows complete obstruction at the level of the eighth dorsal vertebra The superior margin of the opaque column at the site of the obstruction is concave sharply defined and smooth in outline Operation revealed a neurofibroma which had completely occluded the spinal canal

Fig 353 Neoplasm at the level of the fifth to seventh cervical vertebræ on the right side Myelogram There is a large rounded filling defect in the opaque column on the right side at the level of the fifth sixth and seventh cervical vertebræ (black arrow) Operation revealed a neurofibroma

Clinical Aspects The ages in the reported cased cases have ranged from infancy to eighty two years Most of the tumors occur in the third fourth and fifth decades There is a slight predominance in females In many instances the clinical manifestations indicate that the lesion has been present for a period of several months or years prior to the establishment of the diagnosis The commonest symptom in cord tumor is pain and the neurologic disturbances usually coincide with its onset It usually begins in the neck between the shoulders or in the lower back and

bellum In some instances, myelography reveals a persistent obstruction at the level of the first and second cervical vertebrae The head of the pantopaque column assumes a semicircular margin with the concavity directed caudally Turning the patient's head from side to side produces no change in the defect This is due to the downward displacement of the cerebellum The cisterna pontis is narrowed The changes are explained by the fact that normally the inferior aspect of the cerebellum bulges slightly downward and dips into the cisterna magna whereas with the Arnold Chiari malformation, the cerebellum slips downward and forward This distorts its outline and alters the picture in the cisterna magna

The use of air or oxygen as a contrast medium in the study of the upper cervical region may present certain advantages The examination is made with the patient in the recumbent position, sagittal, oblique, and lateral projections being used The subarachnoid space in the cervical region is large and when the fluid in this region is replaced with air there is a sleeve-like collection of gas anteriorly and posteriorly to the cord The position and size of the cord are outlined and abnormalities in the air columns can be demonstrated The cerebellar tonsils are visualized when enlarged and displaced downward, causing a filling defect in the air column In syringomyelia there is enlargement of the spinal cord with narrowing or obliteration of the gas shadows in the subarachnoid spaces The changes are usually less clearly visualized with gas than with opaque media The communication between the subarachnoid spaces and the ventricular system within the cranial cavity normally is widest in the region posterior to the cord as it passes through the foramen magnum At the level of the first and second cervical vertebrae the subarachnoid space is of relatively large volume and in its posterior aspect assumes the shape of an inverted bisected cone The sleeve like collection of gas is best visualized on the oblique or lateral film as an area of diminished density surrounding the cord The anteroposterior roentgenogram is usually not satisfactory because of the density of the vertebral bodies and other superimposed structures Normally the spinal cord passes through the foramen magnum slightly anteriorly to the center of the foramen It is then directed slightly posteriorly and below the level of the second cervical vertebra lies almost symmetrically within the central portion of the canal In the adult the cord has a diameter of about 1 cm at the level of the atlas and axis In the lower segments of the cervical spine there is a gradual increase in size to accommodate the cord and the nerves which supply the upper extremities The enlargement is principally of the transverse diameter of the cord and attains its maximum at the level of the fifth and sixth cervical vertebrae the diameter of the cord at this level being 12 to 13 mm In many instances there is an associated platybasia Spinal cord tumors also may be manifested by defects in the air column or by obliteration of the air column at the site of the neoplasm

ADDITIONAL READING

List C F Neurologic Syndromes Accompanying Developmental Anomalies of the Occipital Bone Atlas and Axis Archives Neurology and Psychiatry *45* 577–616 1941

Marks J H and Livingston K E The Cervical Subarachnoid Space with Special Reference to Syringomyelia and the Arnold Chiari Malformation Radiol *52* 63–68 1949

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It is estimated that 75 to 85 per cent of spinal cord tumors are benign and potentially curable, hence it is of the utmost importance to establish the diagnosis as early as possible and especially before irreversible changes have occurred in the spinal cord. Many of the cases are not diagnosed until late, principally because the lesion had not been considered in differential diagnosis. Tumors of the spinal cord are uncommon. Careful attention to the clinical and roentgen manifestations makes definite diagnosis possible in approximately 95 per cent of the cases.



Fig 352



Fig 353

Fig 352 Neurofibroma. Myelography with pantopaque shows complete obstruction at the level of the eighth dorsal vertebra. The superior margin of the opaque column at the site of the obstruction is concave, sharply defined and smooth in outline. Operation revealed a neurofibroma which had completely occluded the spinal canal.

Fig 353 Neoplasm at the level of the fifth to seventh cervical vertebræ on the right side. Myelogram. There is a large rounded filling defect in the opaque column on the right side at the level of the fifth, sixth and seventh cervical vertebræ (black arrow). Operation revealed a neurofibroma.

Clinical Aspects The ages in the reported cased cases have ranged from infancy to eighty two years. Most of the tumors occur in the third, fourth and fifth decades. There is a slight predominance in females. In many instances the clinical manifestations indicate that the lesion has been present for a period of several months or years prior to the establishment of the diagnosis. The commonest symptom in cord tumor is pain and the neurologic disturbances usually coincide with its onset. It usually begins in the neck, between the shoulders or in the lower back and

tends to radiate along one or both legs or arms In many instances the pain is unilateral and subsequently becomes bilateral Tumors of the spinal cord are apt to have symptoms and neurologic signs referable to all four extremities The pain may radiate around the thorax or abdomen in girdle like distribution Many of the patients sleep in a chair for months because lying down intensifies the pain A common neurologic manifestation is paresthesia, usually consisting of numbness, tingling, and sensations



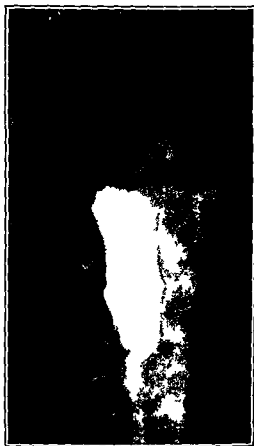


Fig 354 Fig 355

Fig 354 Multiple Myeloma Producing Obstruction and Filling Defects Pantopaque Myelogram The opaque materal was injected in the lower lumbar region With the patient in the Trendelenburg position filling defects were present in the lumbar region on the right The opaque column met with obstruction at the level of the tenth dorsal vertebra The obstruction and filling defects were due to the presence of extradural masses of myeloma

Fig 355 Extradural Meningioma at the Level of the Tenth Dorsal Vertebra Pantopaque Myelogram There is obstruction at the level of the inferior border of the tenth dorsal vertebra At operation a meningioma was found

of heat or cold There is frequently stiffness or weakness of the back, arms or legs and muscle atrophy The deep reflexes of the extremities are increased hypoactive or absent depending on the site and level of the tumor There are disturbances of vibratory and position sense and the sensations of pain heat and cold are altered Intramedullary tumors cannot be differentiated from extramedullary lesions on the basis of the clinical manifestations Many patients have a history of previous operations such as lumbosacral fusions abdominal exploration nephropexy or rib and intercostal nerve resection and the application of body casts



FIG 356 Intramedullary Neoplasm Cervical Region There is an extensive filling defect extending from the level of the third to the sixth cervical vertebrae (black arrow) The defect is intraluminal in character The superior margin of the defect is rounded and presents a cap formation

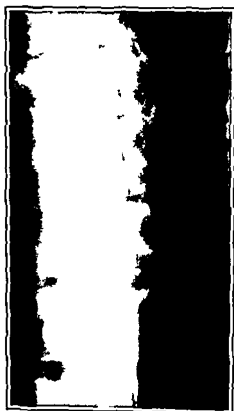


FIG 357 Spinal Cord Tumor with Obstruction at the Level of the Sixth Dorsal Vertebra Pantopaque Myelogram The opaque material meets with complete obstruction at the level of the body of the sixth dorsal vertebra At the site of obstruction there is a rounded defect which is characteristic of intradural tumor Operation revealed an astrocytoma The neoplasm was intramedullary

Laboratory Data The importance of spinal fluid examination in the diagnosis of cord tumor cannot be overemphasized Lumbar puncture should be done at the interspace between the fifth lumbar vertebra and the sacrum since most tumors are cephalad to that level By means of jugular compression and manometric studies of the spinal fluid pressure it may be possible to determine the presence of partial or complete subarachnoid block Observations of the arterial pulsations and respiratory movements in the spinal cord are also important during spinal fluid removal A combination of lumbar and cisternal punctures affords invaluable data Total protein levels exceeding 40 mgms per 100 cc are indicative of



Fig 358 Chordoma Spinogram with Pantopaque Spot roentgenograms made during roentgenoscopy *A* Anteroposterior projection *B* Oblique projection The opaque material was injected into the spine at the level of the second lumbar interspace With the patient in the Trendelenburg position the opaque column meets with complete obstruction at the level of the fourth cervical vertebra The superior margin of the opaque column is rounded and irregular and there is an intralumenal area of radiance at the level of the fifth and sixth cervical vertebræ Operation revealed a neoplasm which on histopathologic study proved to be a chordoma

partial or complete subarachoid block in the absence of degenerative or inflammatory disease of the cord or brain The spinal fluid protein level is usually over 100 mgms per 100 cc and may exceed 1000 mgms per 100 cc While elevation of the spinal fluid protein is not pathognomonic of spinal cord tumor, it is a significant factor in every instance

Roentgen Manifestations Roentgen examination of the spine is of great value in the diagnosis and localization of tumors of the spinal cord Tumors of the spinal cord its coverings and the nerve roots tend to produce local changes in the bones and soft tissues adjacent to the tumor and in consequence it is frequently possible to localize the growth in the spine and show its extent size and type The roentgen changes on the basis

of which a diagnosis of spinal cord tumor may be established are bone destruction, widenings of the spaces between the pedicles and distortions of the paraspinal soft tissues Bone proliferation abnormalities of the intervertebral discs kyphosis scoliosis and abnormal calcifications may also be of significance The commonest and most important finding in the vertebra is bone destruction This is due to pressure of the growth on the adjacent osseous structures with resultant atrophy or absorption As the pressure continues the pedicle or lamina of a single vertebra or the adjacent portions of several vertebra are eroded The destruction may progress to involve the body and the articular processes The change in the pedicles is the most easily recognized and is usually the first to develop A tumor may erode through the intervertebral foramen and cause atrophy of the adjacent portion of the ribs or dislocation of one or more ribs

The Width of the Spinal Interpedicular Spaces

	mm		mm		mm
C 2	30-31	D 3	22-22	D 11	23-27
C 3	30-32	D 4	20-21	D 12	26-30
C 4	32-34	D 5	20-21	L 1	28-33
C 5	32-34	D 6	20-21	L 2	29-33
C 6	32-34	D 7	20-21	L 3	30-35
C 7	31-33	D 8	20-22	L 4	31-35
D 1	27-30	D 9	22-22	L 5	33-39
D 2	23-35	D 10	21-23		

The appearance of the pedicles and the width of the interpedicular spaces varies in the different portions of the spine In the cervical region the pedicles are narrow and the medial borders are flat or slightly convex, while in the thoracic spine, the medial margins of the pedicles are usually convex The pedicles in the lumbar spine appear almost circular At the site of a spinal canal tumor the pedicles may become concave along their mesial aspects In children this is a normal characteristic cephalad of the seventh thoracic vertebra, although in adults this is rarely the case unless disease is present The width of the vertebral canal may be determined with a reasonable degree of accuracy from the roentgen measurements of the distance between the pedicles of the vertebra This is known as the interpedicular distance and is relatively constant in each segment of the spine The measurements may be plotted on a chart and are accurate in patients over twelve years of age In the cervical region the vertebral canal is quite wide The width of the canal decreases rapidly from the first thoracic vertebra to the fifth thoracic level The vertebral canal is fairly constant in width between the sixth and the ninth thoracic vertebrae From the tenth thoracic to the fifth lumbar there is normally a decrease in the size of the interpedicular spaces Tumors of the cord and its coverings frequently produce localized enlargement of the canal hence increase in the width of the interpedicular spaces is an important indication of disease The converse is not true and measurements smaller than the normal are not of pathologic significance If only one interpedicular space is widened, the tumor is probably small When several vertebrae are affected it is an indication that the tumor involves the spine over a corresponding distance

Laboratory Data The importance of spinal fluid examination in the diagnosis of cord tumor cannot be overemphasized Lumbar puncture should be done at the interspace between the fifth lumbar vertebra and the sacrum since most tumors are cephalad to that level By means of jugular compression and manometric studies of the spinal fluid pressure it may be possible to determine the presence of partial or complete subarachnoid block Observations of the arterial pulsations and respiratory movements in the spinal cord are also important during spinal fluid removal A combination of lumbar and cisternal punctures affords invaluable data Total protein levels exceeding 40 mgms per 100 cc are indicative of



Fig 358 Chordoma Spinogram with Pantopaque Spot roentgenograms made during roentgenoscopy *A* Anteroposterior projection *B* Oblique projection The opaque material was injected into the spine at the level of the second lumbar interspace With the patient in the Trendelenburg position the opaque column meets with complete obstruction at the level of the fourth cervical vertebra The superior margin of the opaque column is rounded and irregular and there is an intralumenal area of radiance at the level of the fifth and sixth cervical vertebræ Operation revealed a neoplasm which on histopathologic study proved to be a chordoma

partial or complete subarachoid block in the absence of degenerative or inflammatory disease of the cord or brain The spinal fluid protein level is usually over 100 mgms per 100 cc and may exceed 1000 mgms per 100 cc While elevation of the spinal fluid protein is not pathognomonic of spinal cord tumor it is a significant factor in every instance

Roentgen Manifestations Roentgen examination of the spine is of great value in the diagnosis and localization of tumors of the spinal cord Tumors of the spinal cord its coverings and the nerve roots tend to produce local changes in the bones and soft tissues adjacent to the tumor and in consequence it is frequently possible to localize the growth in the spine and show its extent size and type The roentgen changes on the basis

of which a diagnosis of spinal cord tumor may be established are bone destruction, widenings of the spaces between the pedicles, and distortions of the paraspinal soft tissues. Bone proliferation, abnormalities of the intervertebral discs, kyphosis, scoliosis, and abnormal calcifications may also be of significance. The commonest and most important finding in the vertebrae is bone destruction. This is due to pressure of the growth on the adjacent osseous structures with resultant atrophy or absorption. As the pressure continues, the pedicle or lamina of a single vertebra or the adjacent portions of several vertebrae are eroded. The destruction may progress to involve the body and the articular processes. The change in the pedicles is the most easily recognized and is usually the first to develop. A tumor may erode through the intervertebral foramen and cause atrophy of the adjacent portion of the ribs or dislocation of one or more ribs.

The Width of the Spinal Interpedicular Spaces

	mm		mm		mm
C 2	30–31	D 3	22–22	D 11	23–27
C 3	30–32	D 4	20–21	D 12	26–30
C 4	32–34	D 5	20–21	L 1	28–33
C 5	32–34	D 6	20–21	L 2	29–33
C 6	32–34	D 7	20–21	L 3	30–35
C 7	31–33	D 8	20–22	L 4	31–35
D 1	27–30	D 9	20–22	L 5	33–39
D 2	23–35	D 10	21–23		

The appearance of the pedicles and the width of the interpedicular spaces varies in the different portions of the spine. In the cervical region the pedicles are narrow and the medial borders are flat or slightly convex, while in the thoracic spine the medial margins of the pedicles are usually convex. The pedicles in the lumbar spine appear almost circular. At the site of a spinal canal tumor the pedicles may become concave along their mesial aspects. In children this is a normal characteristic cephalad of the seventh thoracic vertebra, although in adults this is rarely the case unless disease is present. The width of the vertebral canal may be determined with a reasonable degree of accuracy from the roentgen measurements of the distance between the pedicles of the vertebrae. This is known as the interpedicular distance and is relatively constant in each segment of the spine. The measurements may be plotted on a chart and are accurate in patients over twelve years of age. In the cervical region the vertebral canal is quite wide. The width of the canal decreases rapidly from the first thoracic vertebra to the fifth thoracic level. The vertebral canal is fairly constant in width between the sixth and the ninth thoracic vertebrae. From the tenth thoracic to the fifth lumbar there is normally a decrease in the size of the interpedicular spaces. Tumors of the cord and its coverings frequently produce localized enlargement of the canal; hence increase in the width of the interpedicular spaces is an important indication of disease. The converse is not true and measurements smaller than the normal are not of pathologic significance. If only one interpedicular space is widened, the tumor is probably small. When several vertebrae are affected, it is an indication that the tumor involves the spine over a corresponding distance.

Normally, there is a linear area of slight density on either side of the spine which represents the soft tissues of the paraspinal region These densities are best visualized in the thoracic region There is no constant width of the paravertebral soft tissues and no definite alterations occur in relation to age size or sex The margin of this area is normally straight and close to the lateral borders of the vertebræ This is particularly important as displacement or bulging constitutes an important roentgen manifestation of diseases in the region of the spine Tumors of the spinal canal almost never cause bone proliferation The principal exception is hemangioma of the bone and meninges which may produce increased density in the vertebra Tumors such as osteoma osteogenic sarcoma and osteochondroma cause increased density in the bone Arthritic changes of the infectious or hypertrophic type are not significant in the diagnosis of the lesions under discussion The intervertebral discs are rarely involved in spinal cord tumors apparently due to the fact that the cartilaginous tissue is preserved despite pressure on the cartilage or bone Localized narrowing of an intervertebral disc is of pathologic significance Calcification within an intervertebral disc may be important in the diagnosis of cord tumors and has been stated as being a manifestation of chondroma Kyphosis and scoliosis are usually coincidental and are not of diagnostic significance An important exception is in the cervical region kyphosis often occurring in the presence of tumors

Approximately 20 per cent of intradural extramedullary spinal cord tumors cause erosion or infiltration of the spinal column Bone changes occur most often with intradural extramedullary lesions The measurements of the interpedicular spaces are less important than careful study of the pedicles and other bony structures As the tumor enlarges there is progressive erosion of the spinal canal with atrophy of the pedicles, laminæ and posterior surfaces of the vertebræ In the cervical spine, punched out defects of the lateral masses and enlargements of the intervertebral foramens are common The subdural and intramedullary tumors do not usually invade bone although meningiomas may involve several vertebral bodies and the laminæ, pedicles, and spinous processes Multiple erosions of the vertebræ are unusual except in ependymomas, congenital cysts and tumors of the cauda equina The neurofibromas may cause extensive changes in the bodies of the vertebræ Extraspinal extensions of cord tumors may occasionally be demonstrated by roentgen methods the dumb bell type of neurofibroma being the best example of this group The soft tissue extensions are difficult to identify in the cervical spine but are easy to detect in the dorsal spine In the lumbar area they may cause deformation of the psoas muscle shadow

Myelography should not be used until other methods of diagnosis have been exhausted Since delay in the establishment of the diagnosis increases the difficulties of therapy and the gravity of the prognosis, failure to utilize myelography is inexcusable Pantopaque is the best opaque medium Air myelography is not satisfactory in most cases If a subarachnoid block has been demonstrated by a study of spinal fluid dynamics the oil may be injected into the cisterna magna The importance of this lies in the fact that it is helpful to the surgeon to be able to identify the upper level of the lesion in planning the surgical approach The lower limit frequently is determined with reasonable accuracy by the neurologic studies while the upper margins are seldom sharply defined clinically The character of the defect produced by tumors is variable There may

be partial block, widening of the cord, and lateral displacement of the opaque medium. Lateral defects in the opaque column are more often present in tumors which have arisen beneath the dura and extended through the dura into an intervertebral foramen. A subdural tumor which produces complete block is manifested by a central concave defect in the opaque column. With the aid of myelographic examination it is possible to identify and localize approximately 95 per cent of tumors of the spinal cord.

Classification. Spinal cord tumors are classified as extradural, extramedullary and intramedullary. The extradural type is external to the dura and arises in any portion of the spinal canal. They frequently produce changes in one or more of the vertebrae and the surrounding soft tissues. The extramedullary, intradural neoplasms lie in the region between the spinal cord and the dura mater and in many instances can be localized accurately as they are apt to cause changes in the adjacent vertebrae. The intramedullary types arise in the substance of the spinal cord and include the ependymomas, spongioblastomas, astroblastomas, oligodendrogliomas, medulloblastomas, ganglioneuromas, hemangioblastomas, lipomas and neurofibromas or perineural fibroblastomas. It is not always possible by roentgen methods to differentiate the various tumors of the spinal cord or meninges which compress the cord. In some instances, the tumors are of the combined form, being extradural, extramedullary and intradural, and/or intramedullary. Similar changes are produced by space occupying lesions such as gummas, tubercules and parasitic cysts and these must be included in the differential diagnosis.

Extradural Tumors. The extradural neoplasm is located externally to the subarachnoid space. The growth may extend from the meninges through the intervertebral space and form a soft tissue tumor adjacent to the spine. The mass may be round or more commonly long or flat. There is usually no associated erosion of the vertebrae. A round or fusiform soft tissue swelling adjacent to the spine without localized bone destruction is most probably sarcoma or lymphoblastoma. In the cervical region, extradural tumors may cause anterior displacement of the trachea due to infiltration of the growth into the prevertebral soft tissues and produce changes closely similar to those which occur in retropharyngeal abscess. On myelographic examination, there is an indentation of the shadow of the opaque medium with a rounded, sharply defined defect which is characteristic. By rotation of the patient into the oblique and lateral positions, the roentgenoscopist can determine the localization of the maximum area of narrowing. This establishes the anatomic location of the neoplasm. With increase in the size of the tumor, the spinal cord and subarachnoid space are displaced more and more and the defect increases correspondingly. When complete obstruction develops, it is manifested by a block with arrest of the opaque medium at the level of the tumor.

Intradural, Extramedullary Tumors. The meningiomas form about 10 per cent of all the extramedullary spinal neoplasms and are most apt to occur in the thoracic region. They may result in erosion of a portion of one vertebra or parts of several vertebrae. The destruction usually involves the pedicle with unilateral flattening or concavity and widening of the interpedicular space. When calcification is visualized within the spinal cord, it is most often due to meningioma and is an important manifestation which is of invaluable aid in diagnosis and localization.

Both extradural and intradural meningiomas produce essentially similar manifestations. Neurofibromas are also among the common extramedullary tumors. It is important to establish the diagnosis of spinal meningiomas and nerve sheath tumors as early and accurately as possible as most of these neoplasms are potentially curable and can be treated successfully. Practically all of the extramedullary intradural tumors are benign in character. Meningiomas and schwannomas are the most common tumors in this group, primary subarachnoidal and subdural tumors of other types being encountered only very rarely. The meningiomas lie in the intrathecal space, arise from the cells covering the arachnoid villi, and are composed of elongated cells in a whorled arrangement. The schwannomas, also termed neurinomas and neurilemmomas, are nerve sheath tumors and arise from the sheath of Schwann. They consist of cells with elongated nuclei arranged in the form of a palisade. The meningiomas are firm and cause compression and angulation of the spinal cord. While usually subarachnoidal in location, they adhere to the dura over a broad surface. The schwannomas are encapsulated, soft, often cystic and cause a less marked depression in the spinal cord. They are usually attached to the posterior nerve roots, rarely adhere to the dura, are smoother and larger than the meningiomas, and may be multiple. The meningiomas and schwannomas comprise approximately 60 to 70 per cent of the primary tumors of the vertebral canal. They occur with much greater frequency in females and are most common in persons over forty years. They may involve any level of the spine although they appear to be more common in the thoracic region.

Many patients with these lesions do not show any manifestations on the plain roentgenograms. Erosion of the vertebrae or calcification is uncommon and usually occurs only in the advanced cases. The diagnosis can usually be established only by myelography. The procedure is accurate as regards detection and localization and a benign extramedullary, intrathecal tumor should be demonstrated in every case in which it is present. A sharply outlined defect in the advancing head of the radiopaque column comprises the characteristic roentgenologic manifestation. In some instances, estimated at approximately 20 per cent of the cases, it may not be possible to determine whether the lesion is intradural or extradural. Displacement of the spinal cord and spinal nerve roots is more marked in extramedullary lesions. Preoperative definition of the type of tumor is in many instances merely of academic interest as most of the lesions are amenable to complete surgical extirpation. There is partial or complete obstruction to the flow of the opaque medium in practically every case. A meningioma rather than a schwannoma may be suspected when the myelogram discloses a tumor with a broad dural attachment and a slightly irregular surface. The schwannomas are encapsulated as a rule and are surrounded by the radiopaque medium to a greater degree than the meningiomas.

Intramedullary Tumors. The intramedullary tumors comprise about 20 per cent of the intraspinal tumors. The neurofibroma or perineural fibroblastoma is one of the relatively common types of tumor in this group. Neurofibromas may also be extramedullary in origin and both forms produce the same changes in the roentgenogram. It is the tumor which is most apt to give roentgen evidence of its presence and occurs with equal frequency in the various segments of the spine. The most important manifestation is bone destruction. This may vary from slight

erosion of the inner aspect of a single pedicle or lamina to extensive absorption of two three or more vertebrae Pressure atrophy in large neurofibromas in addition to the destruction of the processes and laminae of the vertebrae is associated with extensive concave defects of the lateral margins of the vertebral bodies The tumor may extend through the intervertebral foramen and cause absorption of the proximal portions of one or several ribs The paraspinal soft tissues are distorted the changes varying from a slight bulge to a large mass which extends for a considerable distance laterally to the spine In the thoracic region the tumor is clearly outlined by contrast with the surrounding lung Increase of the interpedicular measurements is commonly present The intradural type may extend through the intervertebral foramen without producing changes in the adjacent bone Intramedullary tumors produce an expansion of the cord and are associated with edema and increased vascularity with enlarged tortuous blood vessels about the cord at the site of the tumor On myelography the expanded cord causes a fusiform defect in the outline of the opaque medium The defect is most marked at the level of the main mass of the tumor and decreases progressively toward the peripheral aspects of the neoplasm The margins of the defect are hazy and poorly defined The dilated, tortuous blood vessels in the region of the tumor may produce filling defects or areas of radiance within the opaque column

Differential Diagnosis The differentiation of intramedullary spinal cord tumors from extramedullary neoplasms is often possible because of certain distinguishing biological characteristics of the intramedullary lesions While extramedullary cord tumors usually extend only a short distance along the spinal cord intramedullary gliomas tend to grow within the cord substance for a considerable length In some cases dilatation of the cord does not produce occlusion of the subarachnoid space in the region of the tumor since the lateral diameter of the spinal cord is slightly greater than the anteroposterior diameter and the subarachnoid space has small out pouchings laterally at *each nerve-root sheath The portions of the* canal which remain patent are the lateral margins Consequently the opaque medium which usually flows along the anterior wall of the canal is displaced laterally at the level of the tumor Since the tumor as a rule extends for a distance of several segments along the cord before producing complete block the opaque medium extends along the lateral margins of the dilated cord and fills out the nerve sheath of each segment The medium may be blocked completely after 3 or 4 segments or may outline the entire extent of the tumor The changes are pathognomonic only in tumors which extend for a distance of several segments along the cord before producing a complete block The myelographic picture is characteristic of tumefaction of the cord and does not necessarily indicate the presence of a neoplasm A cyst of the spinal cord or syringomyelia may produce an exactly similar picture The history and physical findings are essential for accurate diagnosis It is possible to obtain an approximate idea of the extent of the enlargement of the spinal cord by measuring the distance between the streaks of opaque medium Errors are introduced by distortion and magnification, hence the measurements are not entirely accurate and the changes must be interpreted with caution

An aid in diagnosis is the fact that in the intramedullary type of tumor, the appearance at the point of obstruction is that of "capping" the margin of the opaque column usually being curved or lunate Extramedullary lesions as a rule produce a filling defect at the margins of the

opaque column without capping An important characteristic of intra medullary spinal cord tumors is a partial block with lateral displacement of the opaque medium so that it is visualized as a streak or series of streaks along the pedicles of the vertebrae

Calcification in Intraspinal Tumors Abnormal calcification in the region of the skull is of great value to the roentgenologist in the diagnosis of intracranial neoplasms However, it has rarely proven helpful in the diagnosis of intraspinal tumors In the past but little attention has been given to the possibility of the development of calcification within the vertebral canal Dyke states "When a definite irregular mass of lime is seen within the spinal cord, the location and type of tumor has been determined, for such an irregular mass of calcification has always proven to be within a meningioma Brown estimates that 15 per cent of intra spinal meningiomas are of the psammomatous or osteoblastic type and that many of these are demonstrable on the roentgenogram Culver and his associates report five cases of intraspinal meningiomas with sufficient calcification to be visible on roentgenograms of the spine and state that no calcification occurs within intraspinal tumors other than meningiomas The typical meningioma is a discrete encapsulated tumor and is usually oval or spherical in shape The spinal and intracranial meningiomas originate from clusters of arachnoidal cells On histopathologic examina tion the meningioma consists of elongated fibroblastic cells with a whorl arrangement Progressive degeneration with fibrosis hyalinization and calcification of the whorls are common resulting in the formation of psammoma bodies In situations where the psammoma bodies are par ticularly numerous they have a tendency to fuse and form calcium spicules The calcification in spinal cord meningiomas may be of three general types The formation of numerous psammoma bodies results in a homogeneous increase in density of the neoplasm and the tumor is vis ualized as an area of increased density This type of calcification is usually not demonstrable in the sagittal projection, being seen clearly only on the lateral roentgenogram When the psammoma bodies are particularly abundant numerous flecks of calcium are present In psam momas which fuse to form calcium spicules or in the presence of osteo blastic activity within the tumor a discrete irregular mass of calcification forms and is demonstrable on the antero posterior and lateral roent genogram Gray has reported a case showing calcification in a tumor which was found to be a vascular oligodendroglioma or hemangioblastoma Rawling recorded a case of a teratoma containing calcium The roentgen diagnosis of intraspinal meningiomas is uncommon in spite of the fact that a high percentage contain calcium The calcification may not be sufficiently dense to be visible regardless of the quality of the roentgeno gram The shadows of the ribs superimposed on the vertebral canal in the dorsal and upper lumbar regions make visualization of small areas of calcification difficult Tomography and other special roentgen procedures may be of value Very careful search for this type of calcification is essential and will increase the percentage of correct diagnoses

Lipoma

Lipoma within the vertebral canal is very rare The lesion is usually diffuse, infiltrates the roots of the cauda equina and may produce changes very similar to those in other tumors The tumor extends through

the lamina to the subcutaneous tissues and presents as a mass over the sacrum. It is covered by hairy skin and is associated with spina bifida occulta. The lesion may occur as a myelodysplastic process with a single trunk which serves as the point of origin of the lumbar and sacral nerves rather than the normal cauda equina. The anomalous trunks may be the site of lipoma and less commonly other pathologic processes such as dermoid cysts or adhesive bands. The lipoma may be connected with a defect in the spinal lamina and dilatation of the sacral canal usually occurs. There is mild impairment of function of one lower extremity and urinary disturbances. The symptoms begin during early life or in adolescence and increase in severity as the patient reaches adulthood. The late development of symptoms is most probably due to tension placed upon the spinal cord by the greater relative growth of the vertebral column. Relief of symptoms may be obtained by sectioning the adhesion or the mass which binds the conus medullaris to the sacral sac. The lipoma usually involves the leptomeninges in the dura and infiltrates the substance of the spinal cord. The lesion extends over several vertebral segments. It may also occur in the cervical and upper dorsal spine. The outstanding characteristic roentgenologically is extensive bone destruction, a number of pedicles undergoing atrophy due to pressure of the tumor. The pedicles become flat or concave and the medial borders disappear completely. There is marked widening of the spinal canal. The posterior surfaces of the bodies of the vertebra in the involved area show increase of the normal concavity and the ribs may be distorted bilaterally. The interpedicular measurements are markedly increased. The changes may not be uniform, *uninvolved areas alternating with normal segments. The bodies of the* vertebrae appear square in the sagittal projection. The intervertebral discs are usually narrowed.

Ecchondroma

Ecchondromas arise from the posterior margin of the intervertebral discs and are essentially a posterior herniation of the *nucleus pulposus*. They are most common in males of middle age or older. As a *rule* they are not calcified. In the rare instances in which calcification does occur there is a sharply defined area of calcific density measuring 5 mm. to 1 cm in diameter in the vertebral portion of the spinal canal at the level of the *intervertebral* cartilage. The corresponding intervertebral disc may be narrowed and show slight calcific deposit within it. There may be hypertrophic changes about the margins of the involved vertebra.

Von Recklinghausen's Disease (Neurofibromatosis)

Neurofibromas originate in the endoneurium and perineurium, the nerve fibers passing through and around them. The tumors may be intradural, extradural or both. They are most common in the lumbar and cervical areas. The characteristic roentgen manifestation is erosion of one or more pedicles unilaterally. The changes in the vertebrae may be very slight or there may be definite and marked loss of bone. This may involve the lateral masses of one or two vertebrae or the adjacent margins of the ipsilateral pedicles with enlargement of the intervertebral foramen. Large tumors cause displacement of the paraspinal soft tissues on one or both sides. Tumors in the upper dorsal region may encroach upon the apices of the lungs. The retrolaryngeal soft tissue space may be increased in width due to the presence of the tumor.

Teratomatous Tumors of the Spinal Canal

Teratomas are developmental in origin and are comprised of one or more germ layer elements in varying degrees of organization and differentiation They occur only rarely in the spinal canal Extremely slow growth is one of their most characteristic features For this reason the patient may have mild, gradually progressive symptoms for many years in some instances the reported duration of the manifestation being as long as forty years While only about 20 per cent of spinal canal tumors produce alterations on the plain roentgenogram, approximately 50 per cent of the teratomatous tumors show bone changes There is frequently a fusiform enlargement of the spinal canal with a marked increase in the interpedicular distance The pedicles may be decalcified flattened, concave, or eroded Flattening of the pedicles is of importance only when associated with an increase in the interpedicular distance The changes frequently involve several contiguous vertebrae An important manifestation is erosion of the laminae This is difficult to detect in the early stages Erosions of the posterior surfaces of the vertebrae are late manifestations With the use of myelography the presence and level of the tumor can be established with about 90 per cent accuracy

Cauda Equina Tumors

Cauda equina tumors are relatively rare, being much less frequent than ruptured intervertebral discs Tumors of the cauda equina frequently simulate disc lesions and differential diagnosis is difficult or impossible In most instances the diagnosis is established only in the late and hopeless stages About 10 per cent of spinal cord tumors affect the cauda equina Because of the width of the sacral canal neoplasms may be present for considerable periods of time and attain relatively large proportions without causing symptoms Localization may be impossible because lesions at different levels often produce closely similar manifestations Removal of the tumor is difficult due to the fact that several nerve roots pass through or surround the lesion In a series of cases reported from the Mayo Clinic there were fifteen operative procedures for spinal neoplasm and during the same period 100 operations for protruded discs were performed In this series of fifteen cases, 8 of the patients presented a typical disc syndrome The striking similarity of the clinical picture of tumors and ruptured discs results in the orthopedic surgeon frequently treating a tumor of the cauda equina conservatively or by spinal fusion Tumors of the cauda equina have been reported at practically every age period from the teens to the seventh decade of life There is no predilection for either sex

CLASSIFICATION The classification of cauda equina tumors is difficult and preoperative establishment of the type of lesion is usually not possible The malignant neoplasms may comprise the following leiomyosarcoma lymphoma myxofibrosarcoma metastatic carcinoma originating in the thyroid bronchus prostate breast and other areas and neurogenic fibrosarcoma In the benign group the common tumors are the neurilemmoma ependymoma neurofibroma meningioma fibrous cyst ependymal cyst chordoma giant cell tumor and lipoma

SYMPTOMS AND SIGNS The most prominent symptom is pain The pain may be present for long periods before neurological manifestations appear Often the cause of the pain is not established and the diagnosis

of hysteria is made. The pain is usually in the lower extremities and may radiate to the thigh or the perineum. Sciatic radiation is common. In the later stages there is muscular weakness, flaccid paralysis, and impairment of sensation. The advanced stages show involvement of the bladder and rectum and in some instances saddle anesthesia. Lesions at higher levels involve the conus medullaris with the production of severe bladder and rectal changes associated with saddle anesthesia, absence of the ankle jerks, and impairment of the power of erection and ejaculation. Loss of pain and temperature sense are common. Paralysis of the feet is an important manifestation. In most instances the disease has been present for a long time and has progressed to a hopeless stage before the diagnosis is established. An early common symptom is numbness in the legs associated with weakness of the lower extremities. The patient with tumor is less apt to show remissions than are patients with disc lesions. It is not possible on the basis of the physical signs to differentiate between a tumor and a protruded disc.

ROENTGEN MANIFESTATIONS. The roentgen manifestations are an important aid in both the diagnosis and the determination of the level of the lesion. The myelogram is the single most important factor in the establishment of the diagnosis as in many cases the history and physical findings are inconclusive. The myelogram should be utilized in all suspected cases despite the fact that the clinical manifestations may point clearly to a herniated disc. There may be a definite block with a cap formation. An important manifestation is the presence of a filling defect in the opaque column. The defect is usually unilateral and the opaque column is displaced laterally at the level of the lesion. In rare instances the presence of a communicating cyst is shown by the presence of extravasation of the opaque medium into the cyst. In some instances the plain roentgenogram of the spine affords important data. There may be defects in the bones and destruction of the facets or the pedicles of the vertebrae. These findings are not present in the majority of cases.

THE LIMITATIONS OF MYELOGRAPHY

Even in experienced hands, myelography is not an infallible diagnostic measure. In rare instances definite myelographic changes have been noted in cases which subsequently presented negative findings at operation. Certain instances of low back pain have not been cured despite the removal of a protruded intervertebral disc, probably due to protrusions which have recurred or the presence of other protrusions which had not been recognized. In order to be significant, a defect must be constant and preferably demonstrable in more than one view. The size of the defect may not coincide with the size of the herniation because a portion of the lesion may extend beyond the confines of the neural canal. Conversely, the size of the defect may be exaggerated by associated changes in the nerve roots. Since the majority of posterior disc protrusions extend to one side of the mid line, the classical defect is a unilateral indentation at the level of the corresponding intervertebral space. In the lateral view, a unilateral defect may be obscured. Small mid line protrusions may produce so small a central defect that detection is difficult or impossible. A large mid line lesion may obstruct the column partially or completely. Lesions which involve the entire width of the disc produce a bilateral

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Hinkel reports an unusual case in which 3 cc of pantopaque inadvertently entered the venous system of a patient during myelography without serious consequences. The needle was inserted into the subarachnoid space and clear spinal fluid was obtained. The pantopaque was deposited in the neural canal and was noted by the roentgenoscopist to be in the subarachnoid space. It did not immediately enter the vein being observed roentgenoscopically in the spine approximately four minutes after the injection. The patient then coughed and almost immediately the opaque medium was observed to leave the canal by means of multiple curvilinear pathways which represented the veins. The exit was so rapid that in fifteen seconds it was visualized in the inferior vena cava and several tributary veins. It disappeared from these structures completely within about one minute. Apparently the cough was the precipitating factor as breathing, straining and coughing cause a rise of the cerebrospinal fluid pressure to a point many times greater than in the abdominal veins with the patient prone. Abnormal vessels in the cord and meninges are a possible cause. The veins may have been torn or perforated by contact with the needle. There was no objective evidence of embolism or toxic sequelae and no delayed pulmonary complications developed.

NUCLEOGRAPHY DISCOGRAPHY

In the past decade herniations of the intervertebral discs have received increased attention. Operative procedures have been perfected and the operation has been greatly simplified. The surgeon is desirous of exposing as small an area as possible and welcomes any method which permits of more accurate localization of the lesion. Clinically, the localization of the herniation depends upon the history with reference to the distribution of pain and neurological manifestations, especially reflex changes in the extremities. Radiographic evidence, particularly contrast myelography, is an important aid in diagnosis. However, the myelogram shows merely filling defects or indentations involving the dural sac and changes in the nerve roots and sleeve, the prolapsed disc not being demonstrable in actuality. Lindblom devised the supplementary diagnostic method of disc puncture known as discography. His first studies in 1941 and 1944 were anatomic, red lead being injected into the central cavities of the nucleus pulposus. In the presence of radial ruptures in the annulus fibrosus the lead passed along the rupture to the surface of the disc. It was believed that puncture of the disc in the living would cause a lesion. This did not prove to be the case and Lindblom developed the method as a diagnostic test.

Disc puncture requires fluoroscopic control. The insertion of the needle must be accurate to reach the center of the disc. The normal disc is filled by 0.5 cc of contrast medium. Ruptured discs require larger quantities for good visualization. On injection of a disc, pain identical with that originally complained of by the patient may be reproduced. Modification of the technique by Erlacher constitutes an important advance. With his method no opaque fluid finds its way into the dural sac. Disc puncture is not an innocuous procedure and can not be utilized in every case of lower back and sciatic pain. It affords more accurate data than myelography in some cases. The difficulty of the technique precludes discography as a routine measure. The indications for its use are not

defect on the postero anterior view A similar symmetrical defect may be produced by bilateral hypertrophy of the ligamentum flavum An hour glass defect is due to the triangular conformation of the neural canal in the lower lumbar region A unilateral defect with sharply defined upper and lower limits may be produced by unilateral hypertrophy of the ligamentum flavum, anterior bulging of the ligamentum flavum and hypertrophy of the articular facet Edema of the nerve roots may be manifested by defects in the opaque column or obliteration of the shadow of the nerve sheath Asymmetry of the axillary outpouchings may occur under normal conditions

The majority of disc protrusions occur between the fourth and fifth lumbar vertebrae or between the fifth lumbar vertebra and the first sacral segment The first sacral nerve root acquires its sheath between the upper and lower borders of the lumbosacral disc A posterior disc protrusion at this level may compress the first sacral nerve root, yet produce no defect in the opaque column The arachnoid pouchings are clearly defined at the lower end of the neural canal since the nerve roots in this area descend almost vertically Changes caused by the presence of an extradural lesion are not demonstrable in many instances Below the level of the lumbosacral disc interpretation is difficult because of the lumbosacral angulation and sacral tilt which cause the lower portion of the dural sac to be visualized on end with the patient prone or supine Anomalies in the region of the cul de sac are frequent causes of error The cul de sac may be narrowed below the level of the fourth lumbar intervertebral space Smooth, multiple hour glass contractions must be evaluated with care Accentuation of the normal prominences of the lumbar intervertebral discs may cause confusion in diagnosis Irregular, slow movement of the opaque column may be produced by extradural or subdural extension of the contrast medium if the opaque material has been injected into the extra arachnoid area Extradural defects may be produced by degenerative diseases involving the intervertebral discs lesions of the articular facets or apophyseal joints, bony proliferations of the margins of the vertebrae and bulging of the ligamentum flavum

DELETERIOUS EFFECTS OF MYELOGRAPHY

The intrathecal injection of iodized oil may be followed in certain cases by *a mild fever of short duration Less commonly there is headache* and aggravation of the previous symptoms The cells and protein content of the spinal fluid are usually increased and may remain elevated for a period of several days While fatal reactions are rare, they have been recorded Permanent ill effects subsequent to myelography are extremely rare The opaque medium retained in the sheaths of the spinal nerves and the caudal sac may produce a mild arachnoiditis with multiple adhesions between the arachnoid and the dura mater If the dura is opened at operation after the injection the opaque material frequently becomes infected and there is proliferation of the arachnoid membrane Reactions of the pia and arachnoid are less likely if the dura is not opened Many patients in whom the opaque material has not been removed completely subsequent to myelography show small globules of the opaque material within the intracranial cavity None of these patients have complained of symptoms and no permanent ill effects have been recorded

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entirely clear and the dangers from faulty application have not been fully evaluated as yet

Erlacher examined a series of discs removed at autopsy Attempts to stain the nucleus with an aqueous solution of eosin or methylene blue injected directly into the nucleus pulposus resulted in the dye being visualized only in the nucleus pulposus mass After degeneration of the annulus fibrosus, fissures and ruptures directly connected with the nuclear cavity as well as the area of the annulus became stained Large ruptures became filled by the opaque medium The spreading nucleus and its channel of diffusion thus become visible, although the nuclear mass alone is stained Opacification was accomplished with an aqueous solution of 0 4 per cent eosin and 30 per cent potassium iodide This made the impregnation demonstrable by roentgen methods There was complete agreement between the radiographic shadows and the eosin stain, the contours of the stain being identical with those of the nucleus pulposus and the diffusion Deposition of the stain did not occur in the healthy or slightly degenerated annulus fibrosus The actual nuclear contours were visualized and conclusions could be drawn as to the state of degeneration of the disc The disc forms were divided into 5 principal groups by Erlacher (1) The globular nucleus was found mainly in young subjects and indicated a healthy disc without degeneration or herniation (2) The lobed type was the usual form in adults presented a few large lobulations, and herniation or protrusion was unlikely (3) The simple branched nucleus showed a central shadow with a few long narrow branches and indicated a predisposition to herniation Frequently a visible protrusion or a real herniation with free prolapse was demonstrable in this form (4) The multiple branched nucleus with a small central shadow and several branches extending in various directions was considered a transitory form of degeneration and indicated the likelihood of prolapse (5) The spread nucleus presented multiple branches extending in all directions with no single central shadow This represents the typical degenerated disc and is associated with narrowing of the disc In this type herniation must no longer be expected The most significant from the clinical point of view are those with simple branched nucleus (group 3) and the spread nucleus (group 5) The last is manifested on the plain radiograph by narrowing of the disc space A simple branched nucleus occurs with herniations which protrude only slightly over the vertebral body In cases of this type some of the fibers of the annulus fibrosus remain intact and protrude only with pressure In the presence of true herniation (a form of group 3) portions of the nucleus lie outside the limits of the disc and the contrast shadow projects beyond the margins of the vertebral bodies With free prolapse there may be no demonstrable connection with the central shadow the opaque material extending beyond the edges of the disc

Technique Erlacher describes his technique in detail A fine lumbar puncture needle a double cannula at least 12 cm in length an injection syringe and a needle and syringe for procaine anesthesia are necessary A 50 per cent solution of iodine in water is utilized, although the 70 per cent solution is most satisfactory in the case of the fifth lumbar disc The roentgenograms may be taken with the patient lying or sitting The local anesthesia is injected as deeply as possible preferably to the depths of the vertebral bodies in the region adjacent to the spinous processes With the needle in position a radiograph is made for orientation The

lumbar puncture needle is then inserted half an inch lateral to the spinous process and immediately above the superior margin of the spinous process of the adjacent lower vertebra, the needle being directed upward and medially. In this manner, the vertebral foramen is by-passed at its lateral angle and the disc is reached paradurally. In the examination of the lumbosacral region, the needle should be inserted downward and medially, the angle varying with the angle of the disc. The elastic or rubbery resistance of the annulus fibrosus is used as a guide and the annulus fibrosus is pierced. The needle is then inserted an additional 1.5 cm. in order to reach the center of the nucleus. The position of the needle must be checked by antero-posterior and lateral radiographs. The stylet is removed and the opaque fluid injected. About 1 to 2 cc. of fluid is utilized. The injection normally requires considerable pressure. If no resistance is felt, it means that the disc is grossly degenerated or that the needle has been misplaced in the soft tissues. If several discs are to be injected, it is preferable to insert all of the needles first and inject the solution into each with as little delay as possible. Sensations are felt in the affected area by the patient and afford confirmation of the correct position of the needle. As the needle is being withdrawn, it is possible to determine as it passes the spinal canal whether it has pierced the dural sac. After the needle has been withdrawn, the patient performs movements of the trunk to promote uniform distribution of the impregnating fluid. The radiographs are taken rapidly as the opaque agent diffuses in a very short time. The roentgenograms are made in the antero-posterior, lateral and right and left oblique projections with the patient turned to an angle of 35 to 45 degrees.

With this technique, it is possible to remain outside the coverings of the spinal cord and the puncture causes little disturbance to the patient. Undesirable sequels may occur after injection into the spinal theca. Absorption and diffusion result in complete disappearance of the opaque medium within a few hours. When properly performed, none of the opaque fluid enters the dural sac. It has been shown in many thousands of intrathecal injections during myelography that aqueous iodine solutions cause no reactions. The soluble iodine compounds are absorbed from the theca and excreted by the kidney, the urinary tract being visualized on the roentgenogram. Iodine reactions do not occur. This has contributed to the development of myelography with aqueous solutions. In nucleography using the technique described above no injury results from the puncture. It has been suggested on theoretical grounds that the procedure may facilitate herniation of the disc at the point of entry of the needle. Experimental studies show that this does not occur. Nucleography should be restricted to cases in which early operation is contemplated and also those in which diagnosis and localization cannot otherwise be made with accuracy. Myelography permits of general examination of the affected area. Nucleography has the advantages of greater accuracy of interpretation and the demonstration of changes in a particular disc. With nucleography it is possible to show the shape of a disc herniation which, because of its lateral position or small size, may not be demonstrable on the myelogram.

Roentgen Manifestations. The normal disc is visualized as a bilocular collection of dye in approximately the middle third of the disc in the antero-posterior and lateral projections. A degenerated type of disc is characterized by irregular distribution of the opaque material throughout

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Chapter

10

Soft Tissue Roentgenography

Introduction The study of the soft tissues has been largely neglected by many radiologists. Careful observation of these structures is very important and is of inestimable value in the diagnosis of many conditions which otherwise might not be diagnosed. By the use of proper techniques, the normal details and many pathologic conditions of the fleshy structures can be visualized with a sufficient degree of clarity to permit of accurate study. The various components of the body have different degrees of density and therefore absorb x-ray radiation to varying degrees so that it is possible to differentiate them by roentgen methods. In the field of diagnostic roentgenology, there are 5 principal densities (1) gas (2) fatty or adipose tissue (specific gravity of 0.92) (3) soft-tissue structures such as the muscles, nerves, parenchymatous organs and viscera as well as water and the body fluids (4) bone and calcific formations within the body (5) metallic substances introduced into the body by design for purposes of opacification or accidentally as for example, needle fragments, bullets or shrapnel. The iodine compounds are widely used in the study of the urinary tract, bronchial tree and uterus and barium salts are utilized for the examination of the gastro-intestinal tract as these substances offer marked resistance to the passage of the x-rays and are more dense than the structures in the previous groups. The greater the thickness of the tissue, the more radiation it absorbs and the less black it appears on the roentgenogram, so that variations in thickness may be as important as differences in density. Another important factor is the degree of density of the substances or organs adjacent to the region under observation. The air-filled lungs are much more radiant than the heart and great vessels, the roentgen study of the chest being facilitated by the varying radiolucencies in this region. Since the stomach, kidneys, pancreas, adrenals, and gallbladder are of about equal density, plain roentgenograms of the abdomen are of much less diagnostic value and usually require the use of opaque media or gas for accurate delineation.

In the roentgen examination for soft tissue structures it is essential that proper technique be utilized. No single procedure is satisfactory for all soft tissue roentgenograms and only experience, trial and error teach the optimum technique for the individual case. In some instances it is necessary to make several exposures of the same part with different techniques in order to differentiate all of the densities. A fine focus tube and a small cone are helpful. The Bucky diaphragm may be used if the thickness of the region to be examined is sufficiently great. However, in the study of the small type of breast or thinner portions of the extremities, better detail may be obtained without the Bucky. Cardboard holders may give more satisfactory results than intensifying screens in very thin or radiolucent parts of the body. The routine roentgenogram can be utilized for study

practically the entire disc and in some instances beyond the margins of the vertebral bodies apparently due to a rupture of the nucleus pulposus but not of the annulus fibrosus The opaque medium may extend into the spinal canal and beneath the posterior spinal ligaments and often extrudes into the epidural space The opaque column may present a concave deformity caused by pressure of the protruding disc annulus against the dural sac In this type of lesion the injection meets with less resistance and sciatic pain is frequently reproduced by the injection The method does not demonstrate tumors of the cauda equina which may be shown easily by myelography

Discography of dissecting transosseous ruptures of the intervertebral discs in the lumbar region has also proven of value The so called persistent epiphyses of the vertebrae were thought to originate from the marginal epiphyses of the growing vertebral bodies They have been termed persistent limbus vertebrae or intercalary bones and were considered by Schmorl to be fragments of the vertebral margin associated with long standing protrusion of the intervertebral disc as disc tissue and fissures are present in the separation line The persistent epiphyses are usually situated anteriorly but may lie posteriorly Nucleography shows extension of the opaque material from the anterior disc rupture into the epiphyseal line, demonstrating that this line is in reality a fissure continuous with the disc rupture which is dissecting anteriorly

ADDITIONAL READING

Erlacher P R Nucleogrpahy J Bone & Jt Surg *34B* 204 1952

Gardner W J Wise R E Hughes C R O Connell F B and Welford F C X ray Visualization of the Intervertebral Disc A M A Arch Surg *64* 355 1952

Lindblom K Discography of Dissecting Transosseous Ruptures of Intervertebral Discs in the Lumbar Region Acta Rad *36* 12 1951

roentgen film during the time of exposure. Hematomas (Fig. 359) abscesses and soft tissue tumors other than lipomas may be demonstrable as zones of increased density in the soft tissues. After trauma there is edema with thickening and decreased radiance in the soft tissues in the region of the injury. The muscles may show bulging or the lines of differentiation between the various muscular layers may be partially or totally obliterated. A triangular area of increased radiance between the posterior surface of the tibia, the posterosuperior border of the calcaneus and the anterior margins of the Achilles tendon represents an accumulation of fatty tissue. After traumatism to the ankle with resultant edema, this area is replaced by a uniform or slightly mottled shadow which is of the same density as the muscular tissues. Injury to the knee or other articula-

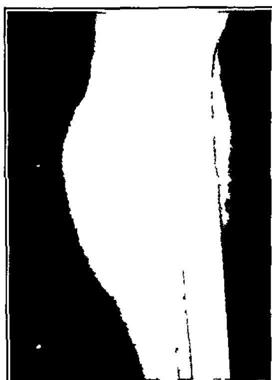


Fig. 359. Hematoma of the Soft Tissues. There is an ovoid area of soft tissue density overlying the upper third of the fibula. The patient had suffered a recent trauma and presented a large hematoma of the soft tissues.

tions may result in increase in the width of the joint space because of the fluid which forms in the joint. The capsule may be visualized as a curvilinear or rounded soft tissue density about the margins of the joint, permitting of a diagnosis of synovitis. This may also occur after infection as infectious synovitis produces a similar appearance. In the case of sprains there is subcutaneous swelling without capsular distensions or evidence of bony injury. Tears of the tendon are common in the quadriceps and Achilles tendons and are manifested by absence of the tendon shadow in the region of the injury with marked swelling and clouding due to edema and hemorrhage. The accumulation of fluid in the bursa may produce a characteristic soft tissue swelling in the region of the joint. This is especially apt to occur in the bursas about the patella and the olecranon process. The distended bursa is manifested by a rounded or ovoid area of uniform density, the density usually being slightly more marked than in the adjacent soft tissues.

of the soft tissues by the use of a strong spotlight or a special lamp with a bulb of 250 watts or stronger A variable resistance and a method of changing the size of the field of illumination are helpful

On the roentgenogram of the extremities, the skin may be visualized because it is outlined between the air on one side and the subcutaneous fat on the other The subcutaneous fatty layers are demonstrable in the extremities, the supraclavicular regions, and along the lateral margins of the torso The individual muscles can be identified in some instances because of differences in thickness In many portions of the body, fat is present in sufficient amounts in the subcutaneous tissues and between the fascia, the muscles and the tendons to serve as a source of contrast which enables the observer to delineate these structures In properly exposed soft tissue films, it should be possible to demonstrate the fibrous strands in the subcutaneous fat and in many instances also the larger blood vessels in the subcutaneous tissues There are particularly large pads of fat in the regions of the knees and the posterior aspect of the ankle which help to outline the tendons, the bursas and the muscular bundles in these regions The subcutaneous fat is relatively large in amount during infancy and in consequence is more clearly visible during this period of life The thickness of the subcutaneous fat increases during the first nine months of life Thereafter it decreases until the thirtieth month This decrease continues at a reduced rate until about the age of five and one half years and at this time it is estimated that the actual thickness averages about half that at the age of nine months In most individuals there is no further change until about the age of eleven or twelve years During puberty there is a definite accumulation of subcutaneous fat this change being more pronounced in girls than in boys

A thorough knowledge of the normal appearances of the soft tissues as visualized on various types of roentgenograms is essential if early or slight changes are to be detected The observer must also understand the location and type of calcific deposits which normally occur within the soft tissues Thus calcification of the costochondral regions or the cartilages of the larynx is not of pathologic significance However, the demonstration of phleboliths in regions of the body where this calcification normally does not occur permits of a diagnosis of venous angioma Faint subperiosteal calcification is the most frequent early manifestation of march fracture and this may not be demonstrable in routine films which have been exposed for bone detail Glass the graphite points of pencils and similar foreign bodies may be obliterated unless special exposures are used Subcutaneous varicosities are often demonstrable in soft tissue roentgenograms In local or generalized hypertrophy of an extremity the soft tissue change may afford very important evidence in determining the extent and type of the condition

In the study of the soft tissue errors in diagnosis may be due to the presence of artefacts clothing dressings and medications on the skin of the patient Tattoo marks while relatively uncommon must also be borne in mind as some of the materials used in the making of tattoos are opaque to the x ray The folds of the skin may enclose air and produce localized increased radiance On the contrary superimposed folds of the skin which do not enclose air cause increased density Superimposition of structures such as the pinna may cause areas of increased density which result in confusion in diagnosis Warts sebaceous cysts and similar soft tissue tumors may be manifested by increased density particularly if impregnated with metallic substances or if pressed tightly against the

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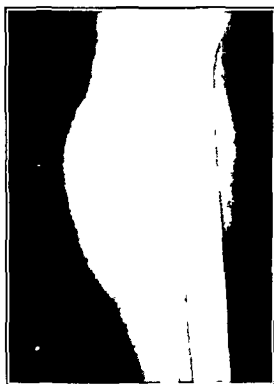


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roentgen film during the time of exposure. Hematomas (Fig. 359), abscesses and soft tissue tumors other than lipomas may be demonstrable as zones of increased density in the soft tissues. After trauma there is edema with thickening and decreased radiance in the soft tissues in the region of the injury. The muscles may show bulging or the lines of differentiation between the various muscular layers may be partially or totally obliterated. A triangular area of increased radiance between the posterior surface of the tibia, the posterosuperior border of the calcaneus and the anterior margins of the Achilles tendon represents an accumulation of fatty tissue. After traumatism to the ankle with resultant edema this area is replaced by a uniform or slightly mottled shadow which is of the same density as the muscular tissues. Injury to the knee or other articulations may result in increase in the width of the joint space because of the fluid which forms in the joint. The capsule may be visualized as a curvilinear or rounded soft tissue density about the margins of the joint, permitting of a diagnosis of synovitis. This may also occur after infection as infectious synovitis produces a similar appearance. In the case of sprains there is subcutaneous swelling without capsular distensions or evidence of bony injury. Tears of the tendon are common in the quadriceps and Achilles tendons and are manifested by absence of the tendon shadow in the region of the injury with marked swelling and clouding due to edema and hemorrhage. The accumulation of fluid in the bursa may produce a characteristic soft tissue swelling in the region of the joint. This is especially apt to occur in the bursas about the patella and the olecranon process. The distended bursa is manifested by a rounded or ovoid area of uniform density, the density usually being slightly more marked than in the adjacent soft tissues.

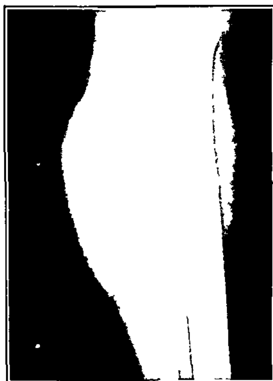


Fig. 359. Hematoma of the Soft Tissues. There is an ovoid area of soft tissue density overlying the upper third of the fibula. The patient had suffered a recent trauma and presented a large hematoma of the soft tissues.

INFLAMMATORY CHANGES

Inflammatory changes produce increase in the thickness of the soft tissue structures Diffuse cellulitis may be diagnosed and its extent determined by the degree of soft tissue change Inflammation in close relation to the subcutaneous fat may extend into the connective tissue reticulum and cause a resultant thickening of the trabeculations Early osteomyelitis causes diffusely increased density in the soft tissues about the involved bone These changes may be demonstrable for a period of one or more days prior to the actual development of periosteal elevation and destructive osseous changes In arthritis there are often swellings of the bursas and increased density of the soft tissues about the involved joints A localized abscess in the muscular layer is usually of the same density as the soft tissues about it and may produce no characteristic roentgen changes If the abscess extends into the subcutaneous fatty layer causes a bulging of the skin, or contains air the presence of the lesion may be demonstrable Displacements of adjacent tissues by an inflammatory mass are of great value in diagnosis An abscess in the lung is visualized because it is more dense than the air filled pulmonary tissue about it An abscess in the liver spleen kidney, or similar organ cannot be differentiated because it is of the same density as the structure within which it lies If there is gas as well as fluid in the abscess, fluid levels may be demonstrable with the patient in the erect or lateral decubitus position In the same manner a brain abscess may not be visualized by routine roentgen methods unless air is present within the abscess Appendiceal, abdominal or pelvic abscess may be diagnosed by displacements of adjacent organs resulting from the abscess The cecum and terminal ileum are apt to be flattened displaced and compressed in appendiceal abscess After the administration of an opaque meal or barium enema these changes as well as fixation of the adjacent organs and localized tenderness on fluoroscopic palpation may permit of a definite diagnosis Pelvic abscess produces similar changes in the bladder, sigmoid and rectum Subphrenic abscess results in elevation and fixation of the diaphragm Perinephritic abscess may be diagnosed with a reasonable degree of certainty by the associated soft tissue changes the psoas muscle shadow and the renal outline being obliterated Retropharyngeal abscess is associated with increased width and density of the soft tissues between the pharynx and cervical spine and anterior displacement and narrowing of the air filled trachea and pharynx Enlarged lymph glands may be demonstrable particularly in the axilla neck and supraclavicular fossa

HEMORRHAGE

Hemorrhage in the soft tissues produces increase in the width and density of the tissues the changes being similar to those occurring in edema Hemorrhage into the joint due to trauma produces widening of the joint space and bulging and thickening of the capsule In hemophilia the presence of blood in the joint produces characteristic changes In the early stages there is widening of the articular space Later there is erosion of the bones forming the joint surfaces with areas of increased radiance in the adjacent bones Hemorrhages may occur in the pelvis or abdomen with displacement and compression of the intestines stomach bladder and other organs At our Clinic there recently was seen a hemo

phaline with severe intra-abdominal hemorrhage. There was a large mass in the left flank. Roentgenograms of the abdomen showed a soft tissue density in the left side of the abdomen. Opaque enema studies revealed marked medialward displacement and compression of the colon by the hemorrhagic mass. Rupture of the spleen may show a characteristic roentgen picture. The shadow of the spleen is obliterated and the stomach is displaced to the right, narrowed, and irregular along its greater curvature aspect; after massive hemorrhage the diaphragm is displaced upward and the splenic flexure and distal transverse colon are depressed. In long standing hemorrhages calcification may occur and hemosiderin is deposited in the soft tissues producing a characteristic mottled granular flocculent density. Subdural hematoma may give no roentgen evidence on the plain roentgenogram. However, pneumoencephalographic studies reveal compressions and displacements of the ventricles. After the lapse of months or years calcification may take place in the hematoma and there may also be changes in the bones of the skull. Myositis ossificans *produces linear or irregular plaque-like areas of calcification in the soft* tissues. The calcific deposits are in some instances in close relation to the bone and usually lie parallel to the planes of the muscles in the affected area.

EDEMA OF THE SOFT TISSUES

Edema may be due to stasis, injury, infection and other causes. The changes in edema are clearly demonstrable on properly exposed roent-

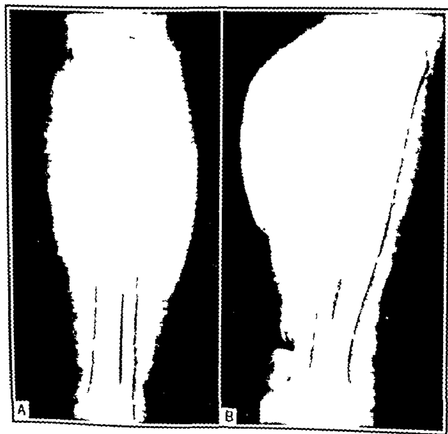


Fig. [illegible]60. Edema of the Lower Leg. *A*, Anteroposterior view. *B*, Lateral view. The lower leg is markedly edematous. There is increased density and swelling of the soft tissues. The septa between the muscles and the differentiation between the muscular and subcutaneous fatty layer are obliterated.

genograms The soft tissue shadows are increased in width and density *The size of a soft tissue mass affected by edema may be considerably* larger than normal The presence of edema may be important in differentiating an inflammatory process from a tumor, as edema occurs commonly in infections but is not seen in association with a neoplasm unless there is obstruction to the lymphatic or venous channels in the region of the growth Roentgenographically, edema is manifested by increase in the thickness and density of the soft tissues In the case of an extremity,



Fig 361 Edema of the Soft Tissues in Erythroblastosis Fetalis There is marked swelling thickening and increased density of the soft tissues indicative of edema

there is an actual increase in the size of the soft tissues, the differentiation of the muscular layers is obliterated and the demarcation of the subcutaneous fatty layer and the musculature is less clearly defined than usual The detail of the bony structures is poor and the roentgenogram appears to have been underexposed or improperly developed In chronic edema of long standing particularly in the case of the lower leg periosteal proliferation may take place with irregularity of outline and widening of the bones Erythroblastosis fetalis produces marked edema of the scalp and extremities (see pp 155–158)

CLUBBED FINGERS

Roentgen studies of clubbed fingers in the past have dealt primarily with changes in the bone. Advanced cases are characterized by increased flaring of the ungual portions of the terminal phalanges. In clubbing which develops during the period of growth the terminal phalanges become hypertrophied and show increase in length. This is particularly apt to occur in the case of unilateral clubbing with venous stasis. In osteitis fibrosa cystica there may occur osteoporosis or complete absorption of several or all of the terminal phalanges. Hypertrophic osteoarthropathy of the bones of the hands with proliferation of subperiosteal new bone is common. In rare cases the terminal phalanges may be encapsulated with newly formed periosteal bone. The etiology of clubbed fingers is unknown. It is believed that the clubbing constitutes hypertrophy and hyperplasia of the tissues about the terminal phalanges as a result of nutritional changes consequent upon increased peripheral blood flow. The soft tissues are increased in thickness and density, the changes being confined to the distal portions of the fingers. All of the digits are affected in equal proportions. No alterations are demonstrable in the bone in the early stages.

MUSCLE ATROPHY

Muscle atrophy occurs in poliomyelitis, certain infections involving the bones and joints, when a limb has been immobilized after a trauma or for other cause, and in the stump of a limb after amputation. The decrease in the size and density of the atrophied muscles can be clearly demonstrated on the roentgenogram. There is also widening of the normal bands which exist between the various muscle groups and of the thin linear shadows which are normally present between the muscle layers. This change is usually best demonstrated by comparison of the two limbs of the same patient. In early tuberculosis of the hip the earliest manifestation demonstrable on the roentgenogram is atrophy of the muscles of the thigh. Careful comparison of the two sides demonstrates this change long before there is actual destruction of the bones about the hip. Similarly in poliomyelitis the degree of wasting of the muscles and the response to therapy may be judged by a study of the soft tissue structures.

MUSCULAR DYSTROPHIES

In primary and secondary myopathy the muscular masses may be altered in size, shape and density. Congenital absence of a muscle or group of muscles is manifested by defects or absence of the shadow of the muscle. In Oppenheim's disease, congenital amyotonia, and Werdnig-Hoffman disease, a primary myopathy, there is a marked decrease in the size of the muscle bundles. The muscles are infiltrated with fatty tissue and there is hypertrophy of the subcutaneous fat. These changes are well shown roentgenographically. Roentgen examination is important in demonstrating the increase in the amount of fatty tissue and the relative decrease in the volume and size of the muscles. Pseudohypertrophic muscular dystrophy in the early stages is characterized by enlargement of the muscular masses. Later large amounts of fat appear in the fascial spaces and in the muscles. Acquired muscular atrophy may produce

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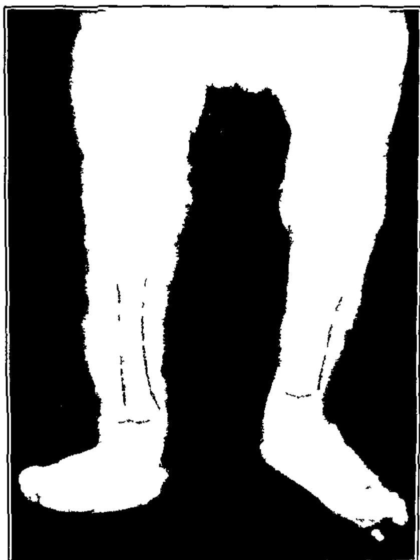


FIG 361 Edema of the Soft Tissues in Erythroblastosis Fetalis There is marked swelling thickening and increased density of the soft tissues indicative of edema

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Fig 363 Fig 364

Fig 363 Fatty Degeneration of the Muscles of the Leg The muscles are thin and wasted There are multiple linear and band like areas of increased radiance extending longitudinally in the muscles indicative of the irregular distribution of fat in the muscles

Fig 364 Muscular Dystrophy There is marked wasting of the muscles and generalized osteoporosis The subcutaneous fatty layer is absent



Fig 365 Arthrogryposis The muscles of the leg are markedly wasted and thin There is increase of the fatty tissue in the subcutaneous regions The patient maintained the legs in flexion and had been unable to walk or stand

shrinkage, fatty infiltration, and fatty degeneration similar to the changes in the primary myopathies

Arthrogryposis Multiplex Congenita or Amyoplasia Congenita This is a rare condition characterized by symmetrical limitation of motion of the joints of the extremities The disease is present at birth and is non progressive in character The deformity is believed to be secondary to an aplasia or dystrophy of the musculature of the extremities as the joints



Fig 362 Pathologic Fracture of the Fibula and Tibia in Anterior Poliomyelitis Marked Atrophy and Fatty Degeneration of the Musculature There is marked decrease in the size of the bones The fractures resulted after a trivial trauma There is marked wasting of the muscles of the thigh leg and foot with areas of increased radiance distributed irregularly throughout the musculature due to fatty degeneration

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Mild trauma resulting from sudden movement such as occurs in paroxysms of coughing may be an important etiologic factor Nontuberculous respiratory disease and rheumatoid arthritis have been present frequently in the disease Dystrophy of the costal cartilages because of malnutrition tuberculosis and trauma do not appear significant in the causation of the condition The second costal cartilage alone is involved in over half of the cases In many instances one or more other cartilages are also involved It has been postulated that microtrauma or involvement by rheumatic disease of the interarticular sternocostal ligament a strong fibrocartilaginous layer which passes within the joint from the second costal cartilage to the fibrous substance between the manubrium and body of the sternum in many individuals although it is not a constant anatomical structure, may be important in the etiology of the disease Dyspnea and cough may produce tears in interarticular sternocostal ligaments with resultant forward rotation of the costal cartilages and predispose to the development of the lesion The clinical manifestations are characteristic Carcinoma of the breast is frequently confused with Tietze s syndrome, resulting in unnecessary operations This is particularly deplorable as the condition is benign and self limited While the pain usually disappears spontaneously in a few days or weeks the swelling may persist for months or years resulting in confusion with metastatic or infectious disease of the ribs Hodgkin s disease and other diseases of grave prognostic import

Roentgen study of the chest ribs and costal cartilages reveals no demonstrable abnormalities Oblique and lateral projections show the localized soft tissue swelling which is palpable and visible on physical examination

CUTIS VERTICIS GYRATA

The term cutis verticis gyrata was applied by Unna in 1907 to an unusual appearance of the scalp characterized by the presence of a variable number of folds imparting to the scalp a superficial resemblance to the gyrations of the brain The scalp becomes thick and may feel oily The changes are most marked in the vertex where the folds frequently extend in an anteroposterior direction, and in the occiput where they tend to lie transversely The changes are demonstrable on roentgen examination Clinical diagnosis is possible only in individuals who are bald since the hair obscures the change The condition may be due to a variety of causes the chief of which are infections of the scalp tumors particularly neurofibromas and certain diseases such as acromegaly myxedema cretinism chronic idiopathic hypertrophic osteoarthropathy tuberous sclerosis Ehlers Danlos syndrome leukemia syphilis and acanthosis nigricans In many cases the changes are idiopathic in character with no associated pathologic condition and are regarded as an isolated developmental anomaly probably an atavism The changes are similar to the arrangement of the scalp in a bulldog It is a dermatologic oddity The patient usually is unaware of its existence Infection may result from accumulation of detritus in the corrugations

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ADDITIONAL READING

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STERN W G Arthrogryposis Multiplex Congenita J A M A *81* 1507 1923

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and is replaced by fibrous tissue. The erythrocytes remaining within the lumens of the blood vessels become concentrated and form agglutinative thrombi of low fibrin content. The thrombi obstruct the vessels and ischemia distal to the site of the lesion results in atrophy and necrotic changes. During the hyperemic stage there is dilatation of the smaller blood vessels and swelling of the endothelium with vacuolization of the musculature of the media. There may be accompanying swelling of axis cylinders of the nerves. During the stage of ischemia the skin shows vesiculation with atrophic changes of the epidermal cells. There is vacuolization and squamous metaplasia of the epidermal cells. In the nerves there may appear degeneration of the axis cylinders, demyelinization accompanied by lipoid phagocytosis and, subsequently, fibrous scarring. The interstitial tissues are atrophic with the development of a gradual diffuse sclerosis which closely resembles post irradiation fibrosis.



Fig. 367 Frostbite. The soft tissues of the great toes are markedly swollen and show irregular mottled density. The changes are more marked on the right. There is osteoporosis of the bones of the feet. The patient had suffered a severe frostbite ten days previously.

The roentgen manifestations in the first days after the freezing comprise marked swelling and increased density of the soft tissues of the affected area. The bones at this time reveal no abnormalities but after a week or ten days show osteoporosis with increased radiance and cortical thinning. Localized areas of necrosis and marked atrophy develop with the passage of time. There is usually no evidence of sequestration or new bone formation. The development of sinuses may result in chronic low grade osteomyelitis with the usual manifestations in the involved bone(s). During childhood the epiphyses are apt to be destroyed with fragmentation, rarefaction and irregularity of outline. The metaphysis adjacent to the affected epiphysis becomes short, flat and irregular in outline with a moth eaten appearance and presents increased density alternating with areas of radiance. The appearance is very similar to that in osteochondritis and may closely simulate Freiberg's disease with which it is frequently confused.

AINHUM

Ainhum is a chronic disease of unknown etiology usually affecting the fifth toe, less commonly the fourth and rarely both. It is characterized by *the spontaneous formation of a constricting ring which appears first on the plantar surface of the toe and finally produces complete encirclement.* As a result of the gradual strangulation there is produced collagenic degeneration of the soft tissues, endarteritis, and rarefying osteitis distal to the point of constriction. The largest number of cases have been reported among the Negroes of the West Coast of Africa, the East Indies and South America. Isolated cases have occurred in Europe, Russia, Istanbul and India. *About forty-five cases have been seen in the United States* principally among the Negroes.



Fig. 366. Ainhum. There is absence of the terminal and middle phalanges and partial absence of the proximal phalanx of the little toe. A linear area of radiance extends across the soft tissues of the little toe, a fibrous band (white arrow). The little toe is markedly decreased in size.

BONE AND SOFT TISSUE CHANGES IN FROSTBITE (PERNIO)

Freezing of a portion of the body results in marked pallor of the affected area. On thawing there is severe hyperemia with redness and heat followed by *swelling and edema.* *In severe cases blebs and vesicles develop* in the involved region. Later there is ischemia, ulceration of the skin and localized necrosis. Stiffness and varying degrees of limitation of mobility ensue. *The pathologic physiology of the changes which develop as the* result of exposure to severe cold is not clearly understood. The initial vasoconstriction of the small vessels after exposure to below freezing temperatures is followed by marked vasodilation. It has been hypothesized that the vasodilation is produced by a histamine-like factor released by the injured cells. Plasma escapes freely through the damaged vascular walls into the interstitial tissues. The plasma tends to organize

and is replaced by fibrous tissue The erythrocytes remaining within the lumens of the blood vessels become concentrated and form agglutinative thrombi of low fibrin content The thrombi obstruct the vessels and ischemia distal to the site of the lesion results in atrophy and necrotic changes During the hyperemic stage, there is dilatation of the smaller blood vessels and swelling of the endothelium with vacuolization of the musculature of the media There may be accompanying swelling of axis cylinders of the nerves During the stage of ischemia, the skin shows vesiculation with atrophic changes of the epidermal cells There is vacuolization and squamous metaplasia of the epidermal cells In the nerves there may appear degeneration of the axis cylinders, demyelinization accompanied by lipoid phagocytosis and, subsequently fibrous scarring The interstitial tissues are atrophic with the development of a gradual, diffuse sclerosis which closely resembles post irradiation fibrosis

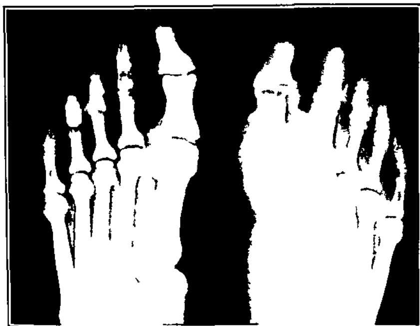


Fig 367 Frostbite The soft tissues of the great toes are markedly swollen and show irregular mottled density The changes are more marked on the right There is osteoporosis of the bones of the feet The patient had suffered a severe frostbite ten days previously

The roentgen manifestations in the first days after the freezing comprise marked swelling and increased density of the soft tissues of the affected area The bones at this time reveal no abnormalities but after a week or ten days show osteoporosis with increased radiance and cortical thinning Localized areas of necrosis and marked atrophy develop with the passage of time There is usually no evidence of sequestration or new bone formation The development of sinuses may result in chronic low grade osteomyelitis with the usual manifestations in the involved bone(s) During childhood the epiphyses are apt to be destroyed with fragmentation rarefaction, and irregularity of outline The metaphysis adjacent to the affected epiphysis becomes short flat and irregular in outline with a moth eaten appearance and presents increased density alternating with areas of radiance The appearance is very similar to that in osteochondritis and may closely simulate Freiberg s disease with which it is frequently confused

TUMORS OF THE SOFT TISSUES

Most tumors of the soft tissues are so similar in density to the tissues within which they lie that accurate differentiation is impossible. However, the neoplasm may be demonstrable and in many instances a correct diagnosis can be established. Roentgen study may make it possible to outline the size, shape and point of origin of the neoplasm. Erosions or invasions of bone and other near-by structures are frequently demonstrable. If the soft tissue mass projects from the body surface, or into the fatty layer, or deforms and displaces surrounding structures, important data with



Fig. [illegible]06. Fibrosarcoma of the Foot. The fifth metatarsal is completely destroyed. There is slight localized destruction of the proximal end of the fourth metatarsal and the inferior surface of the cuboid. The soft tissues of the inferior and external lateral aspect of the foot are markedly thickened and swollen and show discrete and curvilinear areas of calcific density. The bones of the foot and ankle are markedly porotic. A specimen was obtained for histopathologic study and revealed fibrosarcoma.

reference to the lesion is made available. In the regions of the joints soft tissue neoplasms may be visualized as extracapsular or intracapsular masses. The most common periarticular neoplasms are fibromas, fibrosarcomas, xanthomas, synoviomas and angiomas. Cysts of the lateral meniscus of the knee may be demonstrable as sharply outlined prominences at the lateral margins of the joint and differ from synovitis which is characterized by a bilateral bulging with fluid in the suprapatellar pouch. A cyst in the popliteal region is visualized as a rounded area of uniform soft tissue density in the region of the heads of the gastrocnemius muscles. Ganglions, which are most common about the wrist, produce a rounded, localized soft tissue swelling which is slightly denser than the soft tissues about it. Carcinoma of the thyroid may compress or displace the trachea

or extend into the thorax Lipomas and liposarcomas produce a characteristic change in the soft tissues because of the marked radiolucence of the tumors The same may be true of dermoid cysts if they contain large amounts of fat The lipomas are usually subcutaneous and occur most commonly in the region of the neck shoulder breast and thigh Lipomas are also seen in the mesentery mediastinum pericardium lung retroperitoneal tissues and gastro intestinal tract Lipomas of the brain may have a characteristic roentgen picture due to the peculiar butterfly type of calcification associated with the lipoma Lipomas in the subcutaneous fatty tissues of the outer surfaces of the body produce a distinct bulge The margins of the tumor may be outlined sharply by contrast

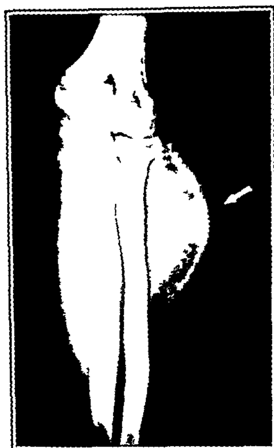


Fig 369 Lipoma The lipoma is visualized as a large ovoid area of increased radiance in the soft tissues of the external lateral aspect of the upper third of the forearm

with the underlying muscles and the skin over it as the lipoma is of greater radiance than the soft tissues within which it lies Some lipomas appear as a mass of bubbles Dermoids in the pelvis and mediastinum may contain large amounts of fatty tissue and be sufficiently radiant to permit of accurate diagnosis from the plain roentgenogram Liposarcoma occurs particularly in the extremities and less commonly in the retroperitoneal tissues of the thorax On the roentgenogram this tumor presents lobulations and irregular mottled zones which are more radiant than normal and through which are scattered areas of soft tissue density due to giant-cells spindle-cells and fibrous strands The dermoids are round or oval sharply defined frequently show a rim of slightly increased density surrounding the area of rarefaction in their central portion and in

many instances contain a formed tooth or bone In the chest, dermoids occur particularly in the anterior mediastinum The lipomas liposarcomas and dermoids may appear more dense than the air-filled lungs when they occur in the chest cavity Confusion with neurofibroma may result in mistaken diagnosis Laryngeal diverticulum may resemble lipoma in some instances Usually, however, lipomas present a very characteristic appearance and diagnosis may be made with a high degree of accuracy from the soft-tissue roentgenograms Large glands, abscesses, and tumors of the neck and axilla are demonstrable and the size and nature of the lesion may be determined in many instances



Fig 370 Lipoma of the Corpus Callosum *A* Lateral view of the skull The lipoma is manifested by an irregular area of increased radiance in the frontal region (arrow) Irregular calcific densities are interspersed within the area of radiance *B* Sagittal projection The lipoma is visualized as a conical area of radiance in the frontal region parasagittally The margins of the lesion are demarcated by a linear irregular area of calcific density (arrows) The roentgen appearances are charactertistic and permit of a definite diagnosis

EPIDERMOID CARCINOMA WITH INVASION OF BONE

Direct invasion of a bone by carcinoma is rare Epidermoid carcinoma involves the bones of the skull maxilla and mandible Squamous carcinoma of the bone originates as a rule from chronic ulcers of the skin or epithelialized fistulous tract The process is most apt to affect the tibia

because the overlying skin is close to the bone. There are two types of invasion carcinoma: ulcer carcinoma in a chronic ulcer and fistula carcinoma which originates in a fistulous tract or a bony sinus. The underlying ulcer or fistula may have been present for years. In consequence the development of carcinoma is not suspected in the early stages at a time when cure may be possible. These tumors are of low grade malignancy and rarely form distant metastases. The diagnosis can usually be established on clinical grounds as there is loss of weight and strength and secondary anemia. In the case of chronic ulcers and fistulous openings there is extension of the lesion and secretion of foul smelling pus. The edges of the wound become indurated, elevated and form cuff like folds. As the tumor



FIG 371 Carcinoma of the Left Antrum. The lateral wall and floor of the left antrum are destroyed and the left naso antral wall is thinned and irregular. There is extensive destruction of the left maxilla. The roentgen diagnosis of carcinoma of the left antrum with extensive bone destruction was confirmed by operation.

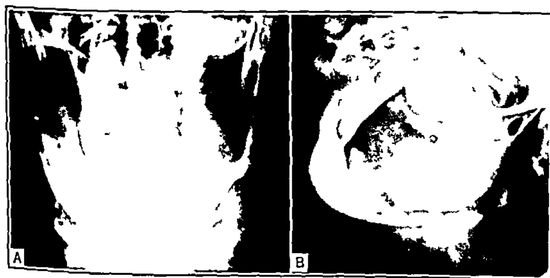


FIG 372 Epidermoid Carcinoma of Soft Tissues with Extension to Mandible. *A* Postero anterior view. *B* Lateral view. There is extensive destruction of the mandible with a pathologic fracture. The fragments show displacement and overlapping. The margins of the area of destruction are markedly irregular in outline. The patient had advanced carcinoma involving the soft palate and tonsillar region with direct invasion of the mandible.

progresses there develop irregular granulations with papillary and wart-like excrescences. A cauliflower type of mass forms. In fistula carcinoma, the tumor may develop near the skin or in the deeper layers near the bone. On roentgen examination there is an area of destruction in the bone. There may be periosteal elevation in the region adjacent to the lesion. Chronic osteomyelitis with eburnation of the bone and sinuses tend to mask early malignant changes. In the later stages, extensive erosion of the bone and pathological fracture occur. Biopsy is essential to establish the diagnosis. Sarcoma presents great difficulty in differential diagnosis. Periostitis secondary to varicose ulcers may lead to an erroneous diagnosis of neoplasm formation. Metastatic lesions seldom develop. With early and prompt treatment a good result may be anticipated. Otherwise, the prognosis is bad and a high mortality results. In the presence of proven carcinoma amputation is recommended. Dissection of the lymph nodes should also be performed.

GLOMUS TUMOR

The normal glomus is a cutaneous structure consisting of an arterio venous fistula without an intermediate capillary bed. The function of the glomus is to regulate the flow of blood to a part of the body. The glomus tumor is a rare benign neoplasm of the skin and subcutaneous tissues arising from the glomus end organ. It usually occurs in the extremities. On histopathologic examination it exhibits considerable variation in structure but resembles the normal glomus and contains the characteristic polygonal epithelioid cells. In some instances the blood vessels in the glomus are large and resemble a true angioma. The lesion is usually small, varying from 5 mm. to 2 cm. in diameter. When located superficially the color varies from deep red to purple or blue and exhibits striking changes in color with change in temperature. The most common location is in the fingers. Exquisite pain occurring spontaneously or after pressure and change in color with temperature change are characteristic. In some instances there are associated sympathetic disturbances such as localized sweating. Most of the lesions are subungual and are associated with a sharply defined area of erosion in the cortex of the terminal phalanx.

PRIMARY CARCINOMA OF THE NAIL

In primary carcinoma of the nail, roentgen examination may reveal swelling of the soft tissues involving the terminal end of the finger and absence of bone at the distal aspect of the terminal phalanx. The extent and site of the destruction are variable. The extreme tuft or the articular portion of the phalanx may be affected. The appearance of the lesion in the sulcus of the nail is the most distinguishing characteristic of carcinoma of the nail. Syphilitic dactylitis, glomus tumor and enchondroma must be considered in differential diagnosis.

ABNORMAL DENSITIES IN THE SOFT TISSUES OF THE PELVIS AND BUTTOCKS

Injections of mercury, bismuth, iodine, iron, and other metallic substances into the gluteal muscles present typical roentgen manifestations. In the days or weeks immediately after injection there form rounded,

droplet like shadows of various sizes and shapes After the lapse of weeks or months roentgen study reveals linear oblique or horizontal bands of increased density in the soft tissues which present a characteristic appearance In rare instances the opaque bands become long and dense and closely resemble ribs the appearance simulating a double exposure with the roentgenogram of the chest superimposed on that of the pelvis



Fig 573 Opaque Material in the Buttocks On the right side the injection had been made about six weeks previously and is beginning to assume a linear pattern The freshly injected opaque material on the left was administered one week previously and is still rounded and sharply defined

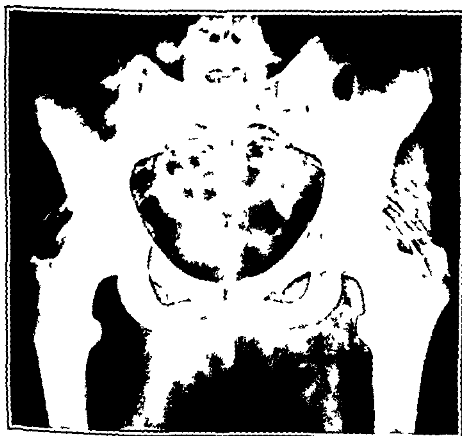


Fig 574 Opaque Material (Bismuth) in the Buttocks The opaque material in the soft tissues has assumed a curvilinear distribution The densities are characteristic and cause no difficulties in diagnosis

Suspensions of bismuth and other metallic compounds represent foreign bodies in the muscular tissues and act as irritants. The material is in large part walled off by connective tissue, although it may be transported in the lymph channels. It is believed that the dense shadows are due mainly to the distribution and retention of the metallic compounds. However, there is probably also an interstitial myositis which produces a degenerative process in the muscle with subsequent deposition of calcium salts. The shadows may remain in the soft tissues for periods of many years depending on the degree of solubility of the material. Follow up studies after three to four years have revealed disappearance of the densities in some instances.

Moles, warts and metallic substances on the skin such as zinc and iodine may produce confusing shadows which are easily recognized by the experienced observer. Lateral views show the exact location and nature of



Fig 375 Soft Tissue Densities Due to Quinine. There are numerous ovoid areas of increased density in the soft tissues of the buttocks. The patient had received quinine injections approximately fifteen years previously.

these shadows. Quinine injections produce a typical ovoid or rounded density which is distinctly different than the changes produced by bismuth and other metallic substances. There may be a trail of density along the track of the needle. Tampons introduced into the vagina cause abnormal shadows in the lower pelvis and abdomen. When the tampons are impregnated with metallic compounds increased density occurs in this region. During menstruation the use of a tampon produces a characteristic appearance of increased radiance in the middle and lower portions of the pelvis.

ABNORMAL GAS SHADOWS IN THE SOFT TISSUES

The presence of gas in the soft tissues or in areas of the body where gas shadows normally do not occur is of great importance in diagnosis. Abnormal gas shadows may be classified as follows: (1) exogenous, which comprises air and other gases which enter the body through a surgical incision, penetrating wound or sinus tract; (2) endogenous, or gas which

escapes from a hollow viscus, and (3) bacteriogenic indicating gas which results from the breakdown of devitalized tissues by the action of bacteria

Exogenous Gas Gas may be introduced during an injury or operation Gas shadows found in or about a superficial wound are usually not of pathologic significance and ordinarily disappear within a few days The gas causes a characteristic area of increased radiance in the soft tissues After operations on the breast air may be trapped in the muscular layers and cause confusion with pulmonary lesions often being considered by the inexperienced observer as due to a fluid level in a hydropneumothorax Fluoroscopy and lateral projections demonstrate that the abnormal shadow is in the anterior chest wall rather than the lung field Gas may be introduced into the abdominal cavity for a diagnostic pneumoperitoneum during laparotomy and peritoneoscopy, or in the treatment of pulmonary tuberculosis and other diseases of the lungs It is essential



Fig 376 Exogenous Gas in the Soft Tissues of the Thigh There are multiple areas of increased radiance in the soft tissues of the thigh characteristic of the presence of gas The patient suffered a compound fracture of the upper end of the femur with fragmentation of the greater trochanter

that the clinician inform the radiologist that an operative procedure has been performed recently as otherwise the patient may erroneously be considered to have a ruptured hollow viscus Trauma to the abdomen the administration of a douche or uterotubography may also result in the introduction of air into the abdominal cavity

Endogenous Gas Gas in the stomach duodenum small bowel and colon normally is recognizable by its characteristic location distribution and shape After perforations gas may escape and usually it accumulates below the diaphragm producing a characteristic appearance which permits of a definite diagnosis Gas may appear in the abdomen after a surgical operation and may persist for two to three weeks Gas forming bacteria within the peritoneal cavity may produce gas If these conditions are excluded it indicates a rupture of the gastrointestinal tract or the bladder As little as 5 cc of air may give a distinct shadow under the diaphragm and 20 cc or more can be definitely recognized

Bacteriogenic Gas Accumulations This type is characterized in the following way (*a*) it appears spontaneously and may not be demonstrable

Suspensions of bismuth and other metallic compounds represent *foreign* bodies in the muscular tissues and act as irritants. The material is in large part walled off by connective tissue, although it may be transported in the lymph channels. *It* is believed that the dense shadows are due mainly to the distribution and retention of the metallic compounds. However, there is probably also an interstitial myositis which produces a degenerative process in the muscle with subsequent deposition of calcium salts. *The shadows may remain in the soft tissues for periods of many* years depending on the degree of solubility of the material. Follow-up studies after three to four years have revealed disappearance of the densities in some instances.

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iodine are used widely in the investigation of draining sinuses of the extremities subsequent to compound fractures the study of abscess cavities the role played by foreign bodies and the study of sequestrums

The injection of sinus tracts and fistulas by roentgen methods may be difficult It is essential that the injection mechanism be tightly applied to the opening of the tract yet the tip be free so that it is not occluded by the underlying structures With a pin point opening a No 20 gauge needle with the shaft cut off near the base is most satisfactory Special conical rubber adapter nozzles may be necessary and can be fashioned from the stoppers used on standard rubber capped vials The stopper is inverted a hole cut in the part which projects into the vial and attached to the metal adaptor of a syringe It may be advisable to utilize the rubber cap of a medicine dropper the open end being cut off and an opening made in the closed end the size of the opening being adapted to the needle base or the syringe Injection through a rubber catheter is often satisfactory A cannula may be threaded through a solid rubber ball and inserted into the tract It is important to utilize gravity or slight pressure in the filling of the sinus tract Excessive pressure must be avoided as the opaque material may enter the veins After injection the opaque material can be retained by having the patient or attendant maintain the syringe in position A piece of gauze may be held over the opening as the syringe is removed Plain roentgenograms should always be made before injection Large films should be utilized in order to outline the entire extent of the tract Stereoscopic roentgenograms are helpful in complicated tracts A film made at right angles to the stereoscopic projection is helpful Oblique views are necessary in most instances although frequently anteroposterior and lateral projections prove sufficient There are no contraindications to the use of iodized oil and no complications ensue It is often possible to demonstrate associated lesions in bone or other structures and foreign bodies whose existence was not suspected Cavities about the joints are always significant and data as to the extent and localization of the cavity is of inestimable value A tract may be long and tortuous and may involve important structures or organs

for several days or weeks after a trauma, (*b*) the location and arrangement of the gas shadows do not correspond to the anatomic boundaries Gas in the peritoneal cavity is confined by the diaphragm or shifts with change in position of the patient to various parts of the peritoneal cavity while gas within an abscess is constantly limited by the margins of the abscess wall, (*c*) there is a tendency to form small, rounded areas of increased radiance, particularly in association with a foreign body, (*d*) successive roentgenograms reveal increase in the amounts of gas the bubbles frequently becoming larger, changing in position, and coalescing, (*e*) fluid levels may appear and are either single or multiple This type of gas develops in association with blood clots or devitalized tissue Abscesses containing gas may be found in the abdomen the pelvis the retroperitoneal region, the subphrenic space, the deep tissue of the axilla, and the liver A common site is in an infected hemothorax In the case of opaque foreign bodies increase in the amount of the gas accumulating about the foreign body is important evidence of infection With nonopaque bodies the gas globules may be of importance in localizing the foreign body The roentgen manifestations vary with the portion of the body in which the gas accumulates In the retroperitoneal region, there occur areas of increased radiance which may outline the margins of the kidney and adrenal More commonly, the shadows are arranged in linear streaks parallel to the course of the muscle fibers The gas may form multiple radiolucent areas of varying size or shape In the thigh and groin, the gas collects along the fascial planes and muscle bundles Intra abdominal accumulations of gas are amorphous or irregular in outline and occur in regions normally not occupied by the gastrointestinal organs Stereoscopic and lateral views are helpful in localizing suspected areas Air in the soft tissues may be an important manifestation of gas bacillus infection and this possibility must always be borne in mind Early diagnosis is essential for the prompt institution of therapy

THE STUDY OF SINUSES AND FISTULAS

A sinus is a tract extending from the skin or mucous membranes to a deep seated focus of suppuration a vestigial structure or an aberrant secreting tissue A fistula is an abnormal tract which communicates from one mucous surface to another, or the skin The walls of chronic sinuses are made up of fibrous tissue and are lined by granulation tissue Long standing sinuses and fistulas may become lined with epithelium from the skin or a mucous surface Persistent tracts are usually due to the presence of sequestra foreign bodies inadequate drainage or epithelization of the tract Obliteration of sinuses and fistulas requires removal of necrotic or foreign material and drainage The use of radiopaque media to demonstrate cavities in the body dates back practically to the earliest days after the discovery of the roentgen rays The method is in widespread use and has become an integral part of roentgen study Suspensions and pastes of heavy metal compounds were at first utilized to demonstrate the presence of sinus tracts and fistulas extending from the external surfaces of the body particularly shallow cavities after wounds of the extremities Iodine compounds have attained wide use in this field of roentgen diagnosis and have replaced other opaque media except barium in the study of the gastro intestinal tract Aqueous and oil preparations containing

as smooth, rounded or oval shadows which measure 2 to 5 mm. in diameter, and are usually multiple rather than solitary. The density is homogenous, although they may present a center of diminished radiance in rare instances. Ureteric calculi may have a very similar appearance and only by urographic methods can a definite diagnosis be made, oblique, lateral, and fluoroscopic roentgenograms with an opaque catheter *in situ* or intravenous urography being necessary to establish their true identity in some cases. Calcifications in the walls of the veins also occur, particularly with varicosities. These are commonly seen in the lower extremities and rarely if ever elsewhere in the body.



FIG. 377. Aortic Aneurysm with Erosion of the Bodies of the Vertebræ. A large aneurysm of the abdominal aorta is faintly outlined by irregular plaques of calcification in its peripheral portion. The anterior aspects of the third and fourth lumbar vertebræ show extensive destruction and erosion due to pressure from the aneurysm (black arrow).

ARTERIOGRAPHY

While the walls of the arteries are frequently sufficiently calcified to be demonstrable, no information as to the functional capacity of the vessel is obtained from the plain roentgenogram. Visualization of the arteries after the injection of an opaque medium is an important aid and may supply data with reference to the degree of involvement and possible means of compensation for arterial occlusions. Various opaque media have been utilized. Diodrast in 35 per cent solution has proven most satisfactory for study of the peripheral circulation and cerebral angiography. For the examination of the heart and great vessels Robb and Steinberg advise the use of 70 per cent solution of Diodrast. Thorotrast has been recommended as it is more radiopaque, however, its use has

THE VASCULAR STRUCTURES

The arteries and veins of the extremities may be visualized on the roentgenogram when they are of large size and lie within the subcutaneous fat tissues, particularly in the popliteal region and similar areas The deeper vessels surrounded by muscles normally are not demonstrable unless calcified Varicosities of the superficial veins may form as worm like tortuous areas of slightly increased density in the fatty layer immediately below the skin Increased density and swelling in relation to enlarged veins may be important evidence in the diagnosis of phlebitis In cases of hypertrophy of an extremity or of a portion of an extremity, soft tissue alterations may give clues as to the cause of the hypertrophy and aid in establishing the diagnosis of an arteriovenous aneurysm, hemangioma lymphangioma or neurofibromatosis Calcific deposits may occur in the walls of the veins in the form of linear bands or plaques Arteriosclerosis with calcification in the walls of the vessels is an important manifestation and calcified arteries are common not only in the extremities but also in the abdomen, chest and skull The pelvic arteries and veins are very frequently calcified and phleboliths in this region are common Within the cranial cavity the internal carotid artery and its branches may be visualized due to calcific deposits in the vessels Intracranial aneurysms in addition to showing calcific deposits cause bone erosion with destructive changes in the sella, the sphenoidal bone or the orbital wall Cutaneous hemangioma of the skin subcutaneous tissue or muscles increases the width and density of the tissues involved and obliterates the normal architecture

CALCIFICATION OF THE ABDOMINAL AND PELVIC BLOOD VESSELS

Calcification of the abdominal aorta and iliac arteries is often obscured by superimposition of the shadows of the spine and sacrum If the aorta is tortuous and dilated the calcific shadows lie adjacent to the spine In the lateral projection the calcification is visualized as mottled or linear densities anterior to the vertebral column Aneurysm formations are frequent and may cause erosion of the lumbar vertebrae Calcifications of the splenic and renal arteries are common particularly in older people Aneurysms in these arteries cause ring like areas of increased density with a radiant area in the center The ring is usually broken at one point particularly in the case of aneurysm of the renal artery The arteries of the pelvis and upper thighs may show advanced calcifications in the walls particularly in advanced age in association with Paget s disease and after the continued administration of vitamin A and D in large doses The individual vessels may be recognized and their courses traced The calcific shadows are easily confused with other abnormalities particularly urinary tract lesions Calcifications of the arteries of the extremities result in characteristic linear or curvilinear densities arranged in a parallel manner which are easily recognized by the trained observer Phleboliths are calcific deposits in organized thrombi usually attached to the walls of the veins in the broad ligament of the uterus the bladder and the prostatic plexus They are most commonly found along the lateral and inferior margins of the pelvis in the course of the lower ureter They appear

rather is a manifestation of increased resistance in the circulatory system distal to the occlusion.

RAYNAUD'S DISEASE. In Raynaud's disease there is spasm of the terminal blood vessels with gangrene, usually symmetrical in distribution, of the terminal phalanges. The condition is most frequently seen in young females, usually during the second and third decades of life. There is a feeling of numbness and cold in the fingers, toes, nose or ears, and pallor of the affected areas. Pain and hyperesthesia are marked. In the later stages, there is cyanosis and lividity, and gangrene may occur. The condition is characterized by a marked degree of atrophy and partial or complete absorption of the distal phalanges. On arteriography, there is diminution of the caliber or absence of filling of the distal portions of the arteries of the fingers.

THROMBO-ANGIITIS OBLITERANS. In thrombo-angiitis obliterans the arteriogram may reveal the coexistence of several phases of the disease, arteries of practically normal appearance occurring in close proximity to those with marked changes. Irregular distribution of the involvement is a striking characteristic of the condition. In the early stages there is slight irregularity of outline and variability in the size of the lumen. Rounded filling defects may occur. Later, the artery is irregular in outline, greatly decreased in size, and tortuous in its course. The arterial channel may be divided and in some areas is almost completely obliterated. In the most advanced stages the arteries become completely blocked. The obstruction may involve any part of the vessel. The point of occlusion is usually rounded with the convexity toward the occluded area.

COLLATERAL CIRCULATION. Collateral circulation is evidenced by the presence of arteries which ordinarily are not visualized roentgenographically. Patients with occlusive arterial disease frequently establish collateral channels which compensate for the occluded arteries and prevent gangrene. The newly developed circulation is characterized by marked tortuosity and variability in the size of the vessels. The arteries are profuse in number and present many anastomoses, whereas normally they are largely longitudinal, pursue a direct course, have few crossings and do not tend to anastomose. With partial obstruction a large collateral branch frequently arises proximal to the point of occlusion, the new artery coursing at right angles to the original vessel for a short distance and then turning to run roughly parallel to the course of the occluded artery. Distal to the site of the lesion it again turns to rejoin the parent vessel. Another method of anastomosis is by lateral branching in which an artery adjacent to the diseased artery sends branches laterally to the site of the affected vessel. The number of collaterals may be large and cross anastomoses are common. A less common development in occlusion is prolongation of normal arteries to replace those which have become obstructed. Thus the interosseous arteries normally do not extend below the wrist. Occlusion of the radial and ulnar arteries results in extension of branches of the interosseous arteries into the hand, explaining why gangrene of the hand almost never occurs.

ARTERIOVENOUS FISTULA. In arteriovenous fistula three findings have been described by Camp and Allen: (1) dilatation of the arteries leading to the fistula, (2) absence of normal filling of the arteries distal to the fistula, and (3) pooling of the arteriographic medium in and about the fistula. There is increased opacity to the roentgen rays in the region of the lesion. This is irregular in degree and extent and has been described as having the

been abandoned because of the late reactions which ensue The drug may be injected by either the closed or open method For the upper arm, the brachial artery is used In the study of the lower extremity, the aorta is injected directly or the femoral artery is entered immediately below the inguinal ligament The films must be made in rapid succession, two or three usually being sufficient It is helpful to make multiple roentgenograms at each examination in order to determine the constancy of the changes and to exclude spasm which is usually transient and causes a temporary decrease in the size of an artery In the case of arteriovenous fistulas, the first x-ray is taken during or immediately after the injection of the opaque medium There is usually neither local nor general reaction after the procedure Extravasation of even large amounts of the opaque material into the soft tissues has produced no permanent damage In the normal arteriogram the vessels are smooth and uninterrupted in contour The lumens of the arteries show a progressive decrease in size Abrupt angulation or sharp changes in caliber are of pathologic significance The method has proven of value in the demonstration of (1) congenital anomalies, (2) alterations in the caliber of the vessels, (3) occlusion, (4) collateral circulations, and (5) certain types of tumors involving the bone or soft tissues

Arteriography in the diagnosis of peripheral disturbances has not come into widespread use because the value of the procedure has not been fully appreciated It is indicated in cases in which clinical diagnosis is uncertain or exact localization of the lesion is desirable A rapid cassette *changer and Fairchild type of angiocardiographic unit afford many ad*vantages but are not essential Roentgenograms of the entire length of the limb are made on two 14 × 17 inch films after the injection of 20 cc of the opaque medium The time interval between the injection and the making of the roentgen exposure is measured accurately with a stop watch and registered, thus permitting of determination of the speed of the circulation Two roentgenograms are made in the anteroposterior and two in the lateral plane with a short interval of relief of the arterial compression between the studies Prolongation of the exposure time may aid in visualization of the vessels It is advisable to use low milliamperage with exposure time of approximately three to four seconds

In the presence of gangrene due to obliterating arteriosclerosis arteriography affords important data as to the optimum site for amputation In aneurysms of the extremities, arteriography demonstrates the size of the lesion and the extent of the collateral circulation Osteomyelitis is associated with an essentially normal circulatory system in the acute stages While the vessels may be slightly dilated, they are arranged in orderly fashion and accumulation of the opaque medium in the region of the lesion does not occur Subacute or chronic osteomyelitis may be associated with arteriolar and venous dilatation and slowing of the circulation Osseous tuberculosis and syphilis and osteoarthritis are characterized by diminution of the blood supply with marked decrease in the rate of blood flow both in the affected and the surrounding tissues Paget s disease and generalized osteitis fibrosa are accompanied by increased vascularity and acceleration of the circulation rate The arteries leading to and extending from an aneurysm are tapered at the peripheral portions of the lesion In thrombosis the point of obstruction is sharply outlined Arteriovenous fistulas may be present in Paget s disease Tortuosity of the collateral vessels is not characteristic of any single disease or group of diseases

rather is a manifestation of increased resistance in the circulatory system distal to the occlusion

RAYNAUD'S DISEASE In Raynaud's disease there is spasm of the terminal blood vessels with gangrene usually symmetrical in distribution of the terminal phalanges The condition is most frequently seen in young females usually during the second and third decades of life There is a feeling of numbness and cold in the fingers, toes nose or ears and pallor of the affected areas Pain and hyperesthesia are marked In the later stages, there is cyanosis and lividity and gangrene may occur The condition is characterized by a marked degree of atrophy and partial or complete absorption of the distal phalanges On arteriography there is diminution of the caliber or absence of filling of the distal portions of the arteries of the fingers

THROMBO ANGIITIS OBLITERANS In thrombo angiitis obliterans the arteriogram may reveal the coexistence of several phases of the disease, arteries of practically normal appearance occurring in close proximity to those with marked changes Irregular distribution of the involvement is a striking characteristic of the condition In the early stages there is slight irregularity of outline and variability in the size of the lumen Rounded filling defects may occur Later the artery is irregular in outline greatly decreased in size and tortuous in its course The arterial channel may be divided and in some areas is almost completely obliterated In the most advanced stages the arteries become completely blocked The obstruction may involve any part of the vessel The point of occlusion is usually rounded with the convexity toward the occluded area

COLLATERAL CIRCULATION Collateral circulation is evidenced by the presence of arteries which ordinarily are not visualized roentgenographically Patients with occlusive arterial disease frequently establish collateral channels which compensate for the occluded arteries and prevent gangrene The newly developed circulation is characterized by marked tortuosity and variability in the size of the vessels The arteries are profuse in number and present many anastomoses, whereas normally they are largely longitudinal pursue a direct course have few crossings and do not tend to anastomose With partial obstruction a large collateral branch frequently arises proximal to the point of occlusion the new artery coursing at right angles to the original vessel for a short distance and then turning to run roughly parallel to the course of the occluded artery Distal to the site of the lesion, it again turns to rejoin the parent vessel Another method of anastomosis is by lateral branching in which an artery adjacent to the diseased artery sends branches laterally to the site of the affected vessel The number of collaterals may be large and cross anastomoses are common A less common development in occlusion is prolongation of normal arteries to replace those which have become obstructed Thus the interosseous arteries normally do not extend below the wrist Occlusion of the radial and ulnar arteries results in extension of branches of the interosseous arteries into the hand, explaining why gangrene of the hand almost never occurs

ARTERIOVENOUS FISTULA In arteriovenous fistula three findings have been described by Camp and Allen (1) dilatation of the arteries leading to the fistula (2) absence of normal filling of the arteries distal to the fistula and (3) pooling of the arteriographic medium in and about the fistula There is increased opacity to the roentgen rays in the region of the lesion This is irregular in degree and extent and has been described as having the

appearance of snow flakes adherent to a window pane In order to demonstrate an arteriovenous fistula, the exposure must be of short duration and should be made immediately after the injection before the opaque material has had an opportunity to diffuse through the veins

ARTERIOSCLEROSIS Arteriosclerotic vessels present a marked narrowing of the lumen and irregularity of outline Complete blockage of the artery due to thrombosis is demonstrable and is of value in determining the level for amputation or other operative procedures Collateral circulation frequently develops

Arteriography in Bone Tumors Denstad reports 2 cases in which arteriography was of great value in the study of neoplasms The first was a malignant melanoma of the thigh Arteriography revealed a soft tissue tumor the size of a plum The femoral artery was displaced laterally in a smooth curve by the mass Within the neoplasm, there was a wealth of vessels which were of even caliber, coarse and tortuous A tumor stain was present with a diffuse shadow of the opaque medium deposited in the very small vessels and capillaries Several minor vessels extended from the tumor to terminate in the region of the cutis as slightly dilated lacunæ The second patient presented a malignant neuroma in the region above the left kidney The tumor was demonstrated clearly by arteriography Some of the vessels were small tortuous and irregular while others were dilated Pope and Seyss performed arteriography in two cases of malignant bone neoplasm, a metastatic carcinoma of the radius and a Ewing's sarcoma of the ulna In the former, there was a blurry shadow *in the region of the defect in the bone* The *tumor itself* was sharply defined and its extension into the soft tissues was clearly demonstrable The intensity of contrast increased toward the peripheral portions of the lesion The retention of the opaque medium within the tumor was prolonged, studies two minutes after the injection revealing multiple mottled areas Histopathologic study post-operatively revealed a rich fibrillary hyalinized connective tissue with very wide blood spaces lined with simple endothelium and filled with cancer cells

Dos Santos reports a comprehensive study of this topic He states that in the study of bone tumors by arteriography it is necessary to use serial arteriograms An arteriogram after injection of the opaque solution into the proximal part of the main artery of a limb affords a picture of the vascular pattern to the smallest branches a few seconds after the beginning of the injection and roentgenograms at intervals of one, two or three seconds supply important data and enable the observer to assess the morphological appearance and study the circulation from a dynamic point of view Observation of the time required for the opaque fluid to pass from the arteries to the veins gives an accurate measure of the speed and activity of the local circulation and demonstrates the extent of communication between the arteriolar and venous network Sarcomas arising in the soft tissues of the thigh present a rich vascular network of hairy appearance arising from the deep femoral artery together with a tangled mass of new vessels most of which are of about the same diameter The irregular network of new vessels extending in all directions is an indication of the uncontrolled growth of the tumor and the changes are typical of the proliferative vascular activity which occurs in neoplasms Inflammatory lesions are associated with simple hyperemia and the arteriolar pattern retains an orderly and regular arrangement with successive branches of gradually decreasing diameter producing a picture of exaggerated cir-

culatory activity rather than one of proliferative new growth Tumors of the sarcoma group may reveal multiple dilatations with the formation of blood pools indicative of neoplasm formation The blood in the presence of a new growth passes almost directly into the veins without the interposition of capillaries This is demonstrated on serial arteriograms by the fact that the veins which emerge from the tumor are visualized at an unusually early stage In the normal twenty-two to twenty four seconds must elapse after the beginning of the injection before the venous system becomes opacified In malignant tumors the passage of blood through the tumor is so rapid that the venous circulation in some instances is clearly visualized within fifteen seconds The increase in the speed of the arterial venous flow is one of the most constant features of the circulation in malignant tumors particularly sarcomas of bone and soft tissue It is the opinion of Dos Santos that the increase in vascular activity is confined to the tumor tissue itself, the venous circulation of normal tissues proximal and distal to the tumor showing no increased speed of filling The first veins to fill arise directly from the tumor itself The abnormal vascular pattern defines clearly the limits of the tumor, data which cannot be obtained from ordinary roentgenograms Arteriographic study of bone tumors outlines the entire abnormal vascular network and makes it possible to determine the degree of invasion of the adjacent tissues Blood pools are particularly apt to occur in osteoclastomas Filling of the venous tree is less rapid than in sarcoma, indicating that the lesion is relatively benign In the use of thorotrast as a contrast medium roentgen study reveals that one or more months after arteriography the tumor remains impregnated with the opaque material This is due to the fact that thorotrast has the property of selective fixation in the reticuloendothelial cells

Benign tumors do not contain new blood vessels and are often relatively avascular The transit time of the contrast material is not altered An important application of arteriography is in the assessment of the response of a tumor to treatment by irradiation The initial arteriogram usually shows the limits of new vessel formation and the richness of the newly formed network and may demonstrate distortion of the main vessel by pressure of the tumor After effective irradiation, the arteriogram presents a more normal appearance The principal vessels are restored to their usual positions and the newly formed vascular network may disappear partially or entirely The changes indicate the degree of vascular sclerosis one of the factors in the determination of the control or regression of the tumor

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Venous Angioma

A characteristic and pathognomonic type of calcification occurs in venous angioma with areas of calcific density deposited in the soft tissues The diagnosis is established by the presence of multiple phleboliths in areas of the body where normally no plexus of veins occurs The phleboliths may be smooth dense calcific shadows with rounded or oval margins

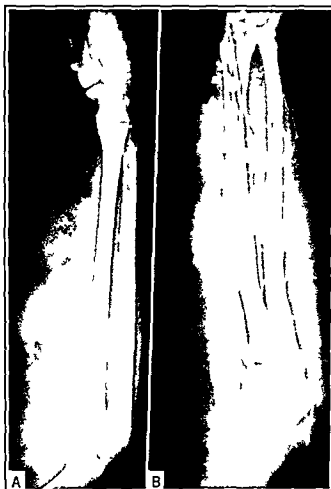


Fig 378 Angioma of Forearm *A* Lateral roentgenogram There is a lobulated swelling in the soft tissues of the anterior aspect of the forearm There are no abnormalities in the bones or joints and no calcific depositions are present in the soft tissues *B* Venogram antero posterior view There are markedly dilated and tortuous veins in the middle and lower portions of the forearm

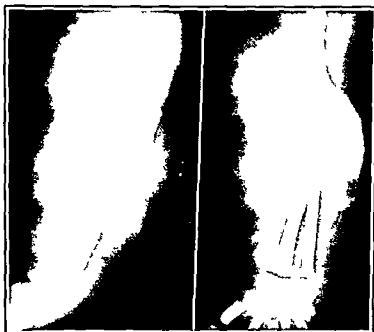


Fig 379 Lymphangioma of Arm The soft tissues of the forearm are very markedly increased in size and density and are irregular in contour The bones reveal no abnormalities The patient was a child six months of age The changes had been present since birth

or may show multiple concentric lamellations This manifestation is pathognomonic and a diagnosis is possible from the roentgenogram Venograms may show a large plexus of veins with venous lacunae The angioma may be visualized as a diffuse density in the soft tissues or as a poorly defined soft tissue mass Therapy usually comprises excision of the mass Radiation may give satisfactory results and may be used when surgery is contraindicated Venous angiomas occur in the extremities the floor of the mouth the liver, the mesentery, intracranially and elsewhere in the body



Fig 380 Venous Angioma There is a large venous angioma of the muscles of the forearm with multiple phleboliths in the soft tissues

Venous Thrombosis

The primary site of spontaneous thrombosis of the leg is usually the deep venous trunks of the calf From this area the thrombus may extend to the popliteal vein and the veins of the thigh Venography may be contraindicated Although injection of the contrast medium does not lead to necrosis as a rule in the presence of acute thrombosis of the lower extremity the vascular supply is compromised and extensive damage may develop Frimann Dahl suggests a method on the basis of which a diagnosis of acute thrombosis may be established from the plain roentgenogram The examination can be performed with a portable x ray unit at the patient s bedside and may be repeated as many times as necessary The casette is placed directly under the calf and thigh and a soft tissue type of exposure is utilized approximately 40 to 50 kilovolts and 50 milliampere seconds at a target film distance of 90 to 100 cm The subcutaneous tissues are clearly visualized with this technique If the thrombosis is believed to involve the upper or middle third of the thigh the film

should include the groin Both sides should be studied for comparison It is essential to include the lateral border of the soft tissues In obese patients a movable Bucky grid is essential In many cases of acute thrombosis the changes may be demonstrable in the soft tissues as early as the first or second day after an attack The roentgen manifestations comprise (1) thickening of the cutis line, (2) accentuation of the reticulated subcutaneous structures due to edema and dilation of the small vessels, (3) widening of the larger vessels, particularly the saphenous veins, (4) increased density of the muscle shadows and obscuration of their margins,

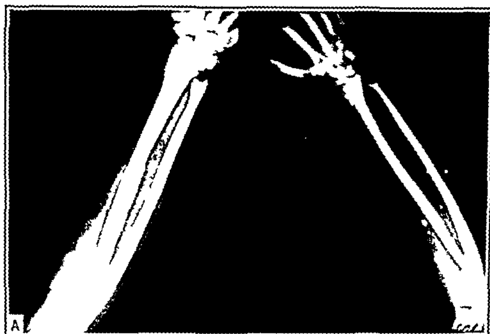




Fig 351 Venous Angiomata of the Left Arm Wrist and Hand *A* There are multiple phleboliths in the soft tissues of the left forearm wrist and hand The left forearm wrist and hand are markedly smaller than the right *B* Photograph shows marked deformity of the left hand and forearm and the decrease *in size of the left limb as compared with the right*

and (5) increase in the width of the extremity. The changes are more clearly demonstrable in patients with abundant subcutaneous fat. They may be present as early as the first or second day. Repeated examinations at intervals of one or two days are essential in doubtful cases. In male patients with poorly developed subcutaneous fat and heavy musculature the only signs may be swelling of the leg and increased density of the muscle shadows. In the late stages there may be widening of the saphenous vein due to occlusion of the deep veins of the calf, the greater portion of the blood stream passing through the superficial veins.

The differential diagnosis must include many conditions. In cardiovascular disease with edema the changes are usually bilateral and are most common in the lower part of the leg and in the region of the ankle. With acute thrombosis the changes in the soft tissues are unilateral, are most common in the widest portion of the calf, and are often best visualized on the lateral aspect rather than along the medial and posterior aspects. The edema and hyperemia of inflammatory lesions can usually be excluded on the basis of the clinical manifestations. In fracture or severe sprain of the leg, edema also is a prominent feature. The history usually establishes the diagnosis. In osteomyelitis changes are present in the soft tissues in the early stages of the disease, often before demonstrable changes are present in the bone. There is accentuation and thickening of the reticulated subcutaneous structures and obliteration of the muscular margins. A similar manifestation may be present in the flank in acute peritonitis and perirenal inflammation.

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Venography

Phlebography has proven of great value in the demonstration of venous obstructions, particularly in thrombo-embolic disease. It is a safe procedure. The widest field of usefulness has been in the demonstration of sites of block in cases of acute post-surgical thrombosis and chronic diseases of the veins. An important physiologic fact that has developed from this work has been the demonstration that the venous return from the legs is much slower than from the arms. Diodrast in 40 to 50 per cent solution is the contrast medium in common use. Venography may be performed by either the indirect or direct method. In the indirect method the opaque material is injected into the artery and flows through the arteries and capillaries into the veins. This method has the advantage of demonstrating most of the veins of the extremity simultaneously. It is difficult to expose the film at the time when the vessels are well filled. Also there is marked dilution of the opaque material so that it may be necessary to repeat the examination several times in order to obtain the desired results. In the direct method, the opaque material is injected into the vein. In the upper extremity, the median basilic, axillary and subclavian veins are clearly visualized. The exact point of injection does not appear to be of significance and any of the vessels of the foot or ankle may be utilized. Twenty cc. of the dye is used, approximately one minute being required for the injection. No tourniquet is necessary. If the leg is

should include the groin. Both sides should be studied for comparison. It *is essential to include the lateral border of the soft tissues.* In obese patients a movable Bucky grid is essential. In many cases of acute thrombosis the changes may be demonstrable in the soft tissues as early as the first or second day after an attack. The roentgen manifestations comprise (1) thickening of the cutis line, (2) accentuation of the reticulated subcutaneous structures due to edema and dilation of the small vessels, (3) widening of the larger vessels, particularly the saphenous veins, (4) increased density of the muscle shadows and obscuration of their margins.

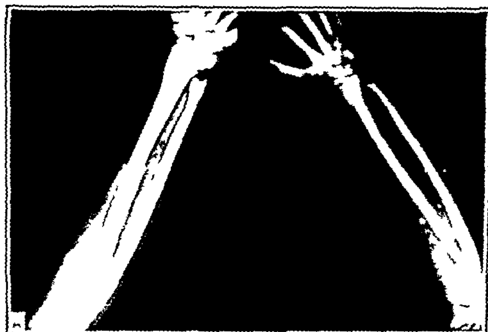




Fig. 3 1. Venous Angioma of the Left Forearm and Hand. There are many phleboliths in the soft tissues of the left forearm, wrist and hand. The left forearm, wrist and hand are markedly smaller than the right. *B.* Photograph shows marked deformity of the left hand and forearm and the decrease in size of the [illegible] as compared with the right.



Fig 384 Venogram Obstruction in the Region of the Proximal Third of the Clavicle The opaque material flows to the level of the proximal third of the clavicle at which point there is complete obstruction There is marked edema of the upper arm with swelling of the soft tissues Mastectomy had been performed for carcinoma of the breast two years previously

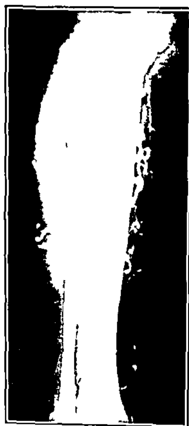


Fig 385 Venogram Varicosities
There is marked dilatation and tortuosity of the superficial veins

swollen and there are large varicosities the dye is injected slowly and films are exposed over a longer period Two 14″ × 17″ films are used and stereoscopic studies are made when possible The leg is maintained in slight inversion for the sagittal roentgenograms One exposure includes the area from the ankle to the knee and another is necessary for the thigh and lower portion of the pelvis The first exposure is made after 10 cc of the contrast medium has been injected, the others following in rapid succession until the completion of the injection The films are placed to overlap each other for several inches so that continuity is obtained

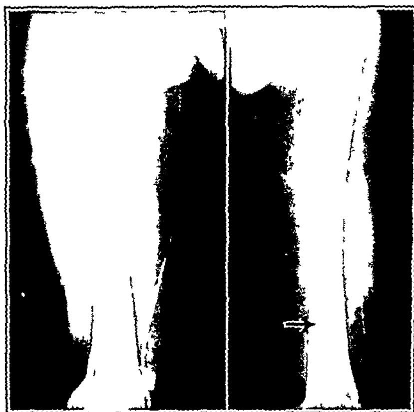


Fig 382 Fig 383

Fig 382 Normal Venogram The femoral vein and the long saphenous vein are visualized and are normal in size and position

Fig 383 Venogram Obstruction of the Femoral Vein There is obstruction of the right femoral vein at the level of the lower third of the femur (arrow)

Lateral views supply important additional data and are made with an additional injection By the use of a 6 or 7 foot target film distance and two screens with Bucky grids it is possible to complete the entire study with one or two exposures

The veins of the lower leg are divided into a superficial and a deep division which communicate with each other through numerous anastomoses The lesser saphenous vein extends from the posterior aspect of the lateral malleous along the posterior surface of the tendon Achilles upward to enter the popliteal vein in the region of the two heads of the gastrocnemius muscle where it anastomoses with the long saphenous vein The long saphenous vein arises anteriorly to the medial malleolus and courses upward along the medial and anterior aspects of the thigh with a



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Fig. 385 Venogram Varicosities
There is marked dilatation and tortuosity of the superficial veins

slight curve at the medial tibial and femoral condyles, to enter the femoral vein at the fossa ovalis There are a few valves, usually 5 or more in number in these veins No arteries accompany them

The deep leg veins are adjacent to the anterior and posterior tibial and peroneal arteries They have numerous valves and lie on either side of the arteries of the leg There are multiple anastomoses between these and also to the superficial veins The anterior tibial vein arises from the deep veins on the back of the foot while the posterior tibial veins originate from the superficial branches of the deep plantar venous system The peroneal vein extends from the dorsal surface of the calcaneus to enter the posterior tibial slightly below the level of the knee The anterior tibial veins lie in front of the interosseous membrane, while the posterior tibials are located in the deep posterior compartment behind the interosseous membrane The popliteal vein is formed by the junction of the anterior and posterior tibials and is adjacent to the popliteal artery The deep veins of the thigh consist of the femoral vein and its branches These lie adjacent to the femoral artery, extending from the popliteal vein to the pelvis to empty into the external iliac vein The femoral vein lies behind and slightly lateral to the femoral artery in its lower portion and medial to the artery in its upper segment

In young people the veins are straight, while in older individuals they are tortuous and dilated In acute superficial block, the medium enters the deep circulation in or slightly above the region of the ankle and the deep circulation is then visualized to the femoral fossa The opaque material flows through the superficial plexus to the point of blockage At this point, the contrast medium stops abruptly or enters a connecting vessel to turn inward or backward toward the deep circulation or to an other portion of the superficial plexus From the point of block upward no superficial plexus is visualized With chronic superficial block, the veins are dilated and tortuous and the upward flow is slow The deep veins fill slowly and show dilatation and tortuosity When a superficial block becomes chronic small connecting veins extend inward toward the deep circulation and as with acute superficial block the medium occasion ally turns abruptly inward toward the deep circulation or toward other areas of superficial plexus In patients with complete block of the entire *deep circulation of the leg and thigh* there may be large dilated super ficial varicosities Thrombi within the veins produce three different types of shadows In one there is a sudden block and the head of the opaque column at this site is concave an indication that the lumen of the vein contains a mass which is outlined by the contrast medium In the second form the vein is not completely obliterated, but contains a ragged, irregular area extending for a variable distance along the wall In the third type the medium follows the wall of the vein and outlines a mass within the lumen Care must be exercised in interpreting single exposures as the findings are often misleading Serial studies are necessary and the constancy of the changes must be established

Non visualization of a portion or all of these veins is common as con genital variations in the pattern are frequently present Incomplete irregular or absent filling of the veins is usually considered pathognomonic of thrombosis or obstruction In the presence of obstruction of the deep vein the superficial veins are dilated and much larger than normal Super ficial channels may be evident in the acute stages of thrombophlebitis and these become more obvious in long standing deep thrombophlebitis A

normal phlebogram can be regarded as definite evidence against thrombophlebitis. However, in view of the fact that so many atypical normal patterns exist, absence of filling does not necessarily mean that a pathologic process is present. It must be stressed that the veins visualized are only those which are open at the time of the injection. In the case of tumors, dilatation, tortuosity, and displacements of the venous structures may be demonstrable. Varices are manifested by increase in the number and size of the veins and the presence of superficial anastomoses.

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Fig. 386. Sialogram. Stone in the Submaxillary Gland. A. Lateral projection. Two small stones are visualized in the region of the angle of the jaw (arrow). There is increased density in the soft tissues surrounding the stones indicative of inflammatory changes in the submaxillary gland. B. Sialogram, lateral projection. The opaque medium outlines a markedly distended duct and delineates the stones in the proximal portion of the duct. C. Sialogram, anteroposterior projection. The gland is well outlined and is large, dense and mottled.

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THE SALIVARY GLANDS

The salivary glands including the parotid, submaxillary, and sublingual, may become enlarged due to inflammation or neoplastic changes Stones in the ducts or glands may be demonstrated on carefully executed roentgenograms, care being necessary to obtain projections in which the calculus is not obscured by the overlying bony densities of the facial bones, hyoid and cervical vertebrae Lateral roentgenograms of the neck and jaw demonstrate a shadow of calcific density slightly below the level of the mandible in the region of the angle or the body in patients with a calculus in the submaxillary gland or the posterior end of Wharton s



Fig 387 Sialogram Parotid Duct The entire duct is visualized The parotid gland is partially opacified by the opaque material

duct The shadow may not be visualized in the sagittal projection because of overlapping of the soft tissue and bony structures of the neck and jaw Stones in the parotid gland are more difficult to demonstrate and require *anteroposterior or oblique projections* Sialography consists of the injection of an opaque medium into the salivary duct To avoid confusing shadows only a single duct should be injected at one time As it may require three days or longer for the opaque material to be eliminated from the gland roentgen investigation of more than one gland should be carried out only after an interval of several days to a week The procedure should not be attempted if there is acute inflammation of the gland In the presence of tumors of the salivary glands there is enlargement with distortion of the glandular structures while extra glandular masses compress and displace the canaliculi Calculi may be localized accurately the sialogram showing whether the stone lies in the duct or within the structure of the gland

CALCIFICATIONS IN THE SOFT TISSUES

Calcifications may be divided into normal or physiologic and pathologic. In the first group are included calcific deposits in the tracheal rings, the pineal body, the costochondral regions and the cartilages of the larynx. Pathologic calcifications occur in practically any tissue and organ of the body. The roentgen examination demonstrates the site, location, and extent of the calcific deposit and in many instances may be of value in determining the underlying etiologic factors. The reasons for the deposition of calcium in various tissues and organs are not clearly understood. One of the most common forms of soft tissue calcification



Fig 388 Fig 389

Fig 388 Dermoid Cyst Containing Dental Structures and Fat There is an irregular area of calcific density lying slightly above the level of the crest of the ilium Partially formed rudimentary teeth are clearly visible in this area The fatty tissue in the tumor is visualized as a rounded area of increased radiance adjacent to the dental structures (arrow)

Fig 389 Calcification in Adenoma of Thyroid Lateral view of the neck. The patient had a large thyroid tumor which had been present for over twenty years There is extensive granular calcification distributed irregularly throughout the mass

occurs in partially or completely devitalized tissue which is not absorbed and becomes infiltrated with calcium salts with the passage of time. Trauma, infection and circulatory changes often appear to play an important role. Disturbances of metabolism frequently result in the formation of calcific deposits. In dead or dying tissue there is decreased carbon dioxide tension because of the lowered metabolic activity. True ossification may subsequently develop in these calcific deposits.

Calcification in peritendinitis occurs most frequently in the tendons inserting in the region of the greater tuberosity of the humerus, the so called subdeltoid or subacromial bursitis. This type of calcification may occur about other joints and has been reported in the region of the hip, elbow, wrist and many other parts of the body. The deposits may be



Fig 390 Calcification of the Testicle *A* Anteroposterior roentgenogram *B* Lateral projection Soft tissue studies of the scrotum show a rounded area of mottled calcification in the region of the right testis The patient had noted a firm non tender mass in this area The testis was removed and on histopathologic examination healed tuberculosis was found



Fig 391 Calcinosis in Association with Edema In association with long standing edema there occurs calcification of superficial tissues The calcification is diffuse and amorphous and is confined to the skin and subcutaneous region

demonstrable two to three weeks after injury and tend to increase progressively in size, in some instances becoming very large. Tumors of the soft tissues may show calcification within the neoplasm. Calcific deposits may occur in primary or metastatic foci and in both benign and malignant tumors. The calcific deposit in many instances results from tissue necrosis due to insufficient blood supply to certain parts of the lesion. The distribution and extent of the calcification vary widely from a diffuse haze which is barely visible to large dense masses which involve the entire growth. In the case of uterine fibroids and ovarian cysts extensive calcification may ensue. Dermoids frequently contain calcific elements and may have well formed dental structures. Osteogenic sarcoma and meningioma often present multiple spicules of bone radiating into the soft tissues, at times in the characteristic sun ray or sunburst formation. Metastatic lesions in the lungs originating in sarcoma of the bone eventually calcify if the rate of growth is very slow. Primary lesions of the pancreas which metastasize to the liver are apt to show irregular calcific deposits within the liver. Myositis ossificans is a deposition of calcium in the tissues and usually occurs after an injury. The calcification begins two to three weeks after the trauma and increases rapidly in size and density. The deposits are usually plaque like or linear, present irregular margins and lie in the muscles and fascial planes. The densities increase in size during a period of weeks or months, the mass becoming more dense with the passage of time.

Moberg described a diffuse type of calcium deposition in the skin and subcutaneous tissues of the legs apparently associated with edema and occurring in individuals with thrombophlebitis, varices and chronic ulceration. The lesions usually are seen in elderly women. The calcification is linear, granular and plaque like and is localized to the superficial tissues of the lower legs. The changes are usually bilateral and extensive, involving the leg from the knee to the ankle. A similar type of calcification occurs in the tissues of the arm although it is rare. Raynaud's disease is characterized by small calcific deposits in the subcutaneous tissues of the fingers. The calcification is due to vasoconstriction and develops in areas of ischemic necrosis. The deposits may occur without sclerodactylia and, while most common in the soft tissues of the fingers, have also been reported about the knee and elbows.

Tuberculosis is one of the common causes of calcific deposits. When it involves the glands it is most commonly seen in the neck and axilla although it may also occur in the groin and the mesenteric glands. The glands tend to lie in chains and extend downward from the neck into the thorax. Healed pulmonary tuberculosis presents extensive calcific deposits in the parenchyma of the lungs. Paravertebral abscess in tuberculosis of the spine is usually calcified about its margins and the course of these abscesses along the psoas sheath is best demonstrated by roentgen studies. Calcification in sebaceous cysts is common.

DERMATOMYOSITIS

Dermatomyositis is a condition in which there is diffuse inflammatory change involving the skin and the underlying muscles associated with fever. The condition is rare and occurs more commonly in children than in adults. The lesion may involve both the muscular tissues and the skin with erythema, edema, induration and swelling. Foci of necrosis in the muscles may undergo calcification and these calcific depositions may persist for many years after the subsidence of the acute process.

CALCINOSIS

A Calcinosis circumscripta
B Calcinosis universalis
Progressive musculocutaneous dystrophy
C Calcinosis associated with excessive ingestion of calcium

Calcinosis is a condition marked by the deposition of calcium in nodules under the skin and in the muscles, tendons, and nerves These calcifications are divided into two groups (1) calcinosis circumscripta in which subcutaneous tissues in the vicinity adjacent to the joints, particularly of the extremities are affected, and (2) calcinosis universalis with diffuse involvement of the deeper subcutaneous tissues and the dermis, usually in the proximal portions of the extremities and the pelvic girdle The frequent association of cutaneous calcifications with vascular disorders of the skin such as scleroderma and Raynaud's disease indicates that the calcific depositions may be related to circulatory disturbances The relationship of calcinosis to scleroderma and other disorders of tissues of mesodermal origin was first shown by Pierge and Wiessenbach in 1905 Until the year 1952 only approximately 150 cases of calcinosis had been described in the literature While the ages of the patients vary widely, calcinosis circumscripta occurs predominantly in persons between thirty-five and fifty five years of age This is in striking contrast to calcinosis universalis which occurs mainly in persons of the younger age groups In the previously recorded cases approximately 30 to 40 per cent showed evidence of scleroderma Calcinosis is considered a manifestation of a degenerative disease in tissues of mesodermal origin On histopathologic study of the tissues, there is found perivascular infiltration of mononuclear cells obliterative endarteritis and hypertrophy of the collagenous connective tissue These changes are associated with generalized fibrosis impairment of the blood supply and subsequent necrosis Areas of fibrosis occur in association with the depositions of calcium salts Degenerative changes occur in fibrous and fatty tissue before calcium deposition ensues Peripheral vasomotor disorders and Raynaud s disease are commonly associated with calcinosis and syphilis and malaria have been precursors in some cases The skin over the calcium nodules may ulcerate and a cheesy purulent material exude The depositions consist mainly of calcium phosphates and carbonates and are often associated with giant cell formation and inflammatory reaction In many cases the calcium content of the blood is high, while in others it is depressed or normal The level of the phosphates in the blood stream may be increased It is the general consensus that calcium metabolism is not altered

A **Calcinosis Circumscripta** Calcinosis circumscripta usually affects the terminal phalanges of the fingers and the extensor aspect of the elbows and knees In these regions large fluctuant masses may be present The deposits are multiple and tend to be symmetrical Females are more frequently affected than males The roentgen diagnosis is facilitated by the fact that the calcium deposits in the skin and subjacent tissues are discrete and mottled and present a characteristic appearance The depositions usually occur earliest in the skin of the digits and tend to be found at the pressure points along the flexor surfaces of the skin at the tips of the fingers The condition is differentiated from myositis ossificans in that in the latter there is plaque like formation of new bone In hyperparathyroidism there is marked osteoporosis of the skeleton and deposition of calcium in the respiratory and urinary systems



Fig 392 Calcinosis *A* Anteroposterior view *B* Lateral view There are numerous small irregular areas of calcific density in the soft tissues of the anterior aspect of the knee The calcific deposits are in the subcutaneous fatty layer There are no abnormalities in the bones or the joint



Fig 393 Calcinosis Universalis There is atrophy of the muscles and the subcutaneous fatty tissues Diffuse granular calcifications are distributed irregularly in the superficial layers of the soft tissues There is marked generalized osteoporosis

B **Calcinosis Universalis** In calcinosis universalis, there is diffuse and extensive calcification of the skin, subcutaneous tissues, tendons, fascia, muscles, and along the nerves The condition occurs in young persons, more commonly in females and does not appear to be associated with tissue necrosis Calcinosis universalis has been recorded at the age of five weeks It is often associated with scleroderma and sclerodactylia The flexor aspects of the limbs are more prone to be affected and the fingers usually escape The joints may become swollen and stiff The serum calcium and phosphorus are usually normal There is apparently an increase in the ability of the body to retain calcium and phosphorus associated with an abnormal affinity of the soft tissues for calcium Mottled and granular calcific depositions are widely distributed throughout the soft tissues of the body in large plaques confined to the subcutaneous regions In the late stages, the patient becomes emaciated there is marked atrophy of the muscles, and decalcification of the bones Draining sinuses form when the skin overlying the calcific deposition breaks down There is pain and stiffness of the involved area with resultant disability Fatal termination is common

WERNER S SYNDROME PROGRESSIVE MUSCULOCUTANEOUS DYSTROPHY In 1934 Oppenheimer and Kugel reported for the first time in the United States a heredofamilial disorder characterized by scleroderma, bilateral juvenile cataracts precocious greying of the hair and other endocrine stigmata which they termed Werner s syndrome The condition is rare and only a few cases have been recorded in the American literature All of these showed remarkable similarity in clinical and roentgen manifestations The etiology of the disease is obscure There is no doubt that it is familial The disease is essentially a degenerative process and appears to be caused by a defect in the mesodermal and ectodermal tissues The syndrome is characterized by premature senescence associated with some features of juvenilism There is generalized underdevelopment of all the endocrine glands resulting in hypogenitalism gynecomastia and eunuchoidism In the female there are menstrual disorders including premature total amenorrhea which may develop as early as the age of sixteen years In some cases there is evidence of thyroid insufficiency Vascular changes *are an outstanding feature of the disease There is clinical evidence of* extensive and widespread arteriosclerosis of the Monckeberg type which may begin in the third decade of life The peripheral vessels are tortuous and cannot be compressed The major changes occur in the ectodermal structures It is not known whether these are secondary to vascular impairments or are primary in nature The predominant changes in the skin are a combination of scleroderma and poikiloderma These are most marked in the distal portions of the extremities particularly in the lower limbs and often are associated with ulcer formations and subsequent gangrene which may require amputation All cases show prematurely early grey hair with the onset as early as the eighth year The greying is rapidly followed by alopecia Bilateral cataracts are constantly present generally begin in the third decade, and progress rapidly so that early in life these patients require surgical intervention to prevent total blindness The voice changes in character usually being described as rasping and high pitched There has been little contributed by pathological studies The afflicted members of the family look very much alike and are frequently mistaken for one another They usually die in the early forties The causes of death are cardiac failure, advanced coronary disease pri

mary carcinoma of the liver, and fibrosarcoma with metastases. In one patient there was evidence of increased parathyroid activity. In a case described by Hershstone and Bower there was a positive Hamilton test and the blood calcium levels were elevated, indicating hyperparathyroidism. The other endocrine glands are usually small and atrophic.





Fig 394 Calcinosis Interstitialis Universalis *A* Pelvis *B* Elbows

Roentgen Manifestations The roentgen findings are those of extensive calcification of the arteries and of the soft tissues The arterial calcification is of the Monckeberg type and involves all the vessels except those of the brain and lung The changes are most marked in the distal parts of the extremities particularly the lower extremities It is possible to identify all the arteries including the plantars and dorsals as well as their branches There is extensive calcification of the abdominal vessels particularly in the mesentery The calcification of the soft tissues is most marked in the subcutaneous tissues, the ligaments, the tendons and the bursas and at times is so extensive that it is not possible to tell which structures are involved There is calcification of the tendon of Achilles and about the calcaneus The knee joint shows widespread involvement with calcification of the medial and lateral ligaments the patellar ligaments and other ligaments and tendons There may also be involvement in the region of the hip in the trochanteric bursa, and in the soft tissues about the interphalangeal joints The depositions vary widely in size In many instances the sella is small The larynx may show advanced calcification of the cartilages There is marked atrophy of the lower extremities best explained on the basis of disuse rather than because of generalized osteoporosis Nodules occur subcutaneously, probably due to the overlying tissues becoming secondarily affected with chronic ulceration

C **Calcinosis Associated with Ingestion of Excessive Amounts of Calcium** Soft tissue swellings which on roentgen examination contain calcium and extensive calcific depositions in the walls of the arteries may occur after prolonged ingestion of alkaline hydroxide gel and alkaline powders in the therapy of peptic ulcer Blood studies reveal a high level of serum calcium Therapy consists of elimination of calcium containing foods from the diet and withholding alkalies except aluminum hydroxide The sequence of events is similar to that in gout The capacity for the retention of calcium in soluble form is exceeded and calcium deposits in insoluble form The long continued and excessive ingestion of the alkali probably is responsible for the initiation of renal damage As the syndrome has been noted after the drinking of enormous amounts of milk, it has been termed milk poisoning

Fibrositis of the Penis Peyronie's Disease

Peyronie s disease is a fibrous cavernositis or sclerosis and comprises a plastic induration of the penis of unknown etiology It is not uncommon and has been described in patients from twenty two to eighty four years of age although the majority of cases occur in the fifth and sixth decades A faulty connective tissue metabolism appears to play an important etiologic role The factor of trauma cannot be minimized The disease has been found after gonorrhea chancre a blow or other injury a break during erection in sleep or otherwise urethral manipulation, stricture of the urethra and surgical operation with resultant blood clot The condition causes pain and distortion The induration appears as a plaque nodule or cord and is solitary or multiple It is most commonly seen in the dorsum in the mid line near the base and may involve the sheath of the *corpora cavernosa* and the *septum* The urethra and the *corpus spongiosum* are not affected The induration is superficial and readily palpable The skin over the area is movable and not tender Micro

scopically the lesion resembles a keloid consisting of a network of scar like and elastic tissue with few vessels. When the disease has been present for a considerable period of time deposition of bone and cartilage occurs. The calcific deposits may be round, oval, irregular or plaque like and vary in size from a few millimeters to several centimeters in extent. Radiation therapy affords relief and practically all forms of roentgen and radium treatments have proven effective. Sterility and loss of libido may result and these therapeutic measures must be applied with caution.

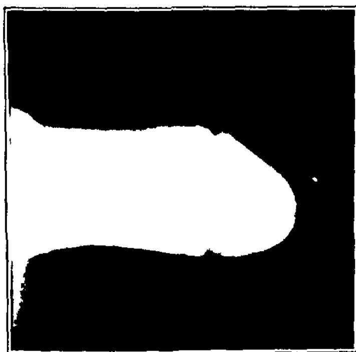


Fig. 395 Peyronie's Disease. There are multiple linear and irregular plaques of calcification distributed irregularly throughout the soft tissues of the penis.

Calcification in Echinococcus, Filaria, and Cysticercus Disease

Echinococcus cysts of the liver and lungs are very prone to calcify. The cysts occur less commonly in the muscles. The calcification is usually in the form of a thin rim about the wall of the cyst. The plaques are curvilinear in shape and may outline the entire margin of the mass. The roentgen shadows are usually mottled and irregular as the density of the calcific plaques varies widely. The trichinella spiralis is very prone to calcify. This parasite frequently lodges in the diaphragm and intercostal muscle. However the larvae are usually too small to be demonstrable roentgenographically. The guinea worm is rarely encountered in the United States but is very common in other countries. While most frequently seen in the subcutaneous tissues of the lower extremities, it may occur anywhere in the body. The calcified worm may be detected as a linear or curved mass. Calcified worms of other types, particularly the filaria bancrofti may be demonstrable on roentgen examination. The calcific deposits are usually in the subcutaneous tissues, the lymph nodes, and the lymphatics of the scrotum. They produce small elongated shadows of density usually closely simulating phleboliths. Calcified guinea worms may attain a length of several centimeters and may assume

the form of segmented chains, coils or lines, although most commonly the densities are about the size of rice grains They may occur anywhere in the body including the peritoneum

Dombrovsky during a roentgen examination of a lumbar spine discovered along each side of the spine a series of dense shadows with sharply outlined borders The shadows each measured about 9 mm × 2 mm, were spindle shaped, and the longitudinal axis corresponded to that of the muscle fibers They were considered to be calcified parasites located in the muscles Further studies of the patient revealed similar inclusions in the soft tissues of the thighs, legs, upper arms, forearms, hands and thoracic wall Biopsy performed on a specimen of the tissue revealed a calcareous deposit with a diameter of about 3 mm which on microscopic examination showed a partially calcified cysticercus The cysticercus represents the larval stage of the band worm called Taenia solium Man



FIG 396 Calcified Worms in the Soft Tissues The soft tissues of the thighs contain multiple small linear and punctate areas of increased density The shadows were discovered during a routine roentgen study after a trauma On excision the calcific deposits were found to be in worms tinea saginata

is the constant host and is usually the bearer of the adult worm only the latter living in the human intestines The eggs of the band worm are excreted in the feces On penetration into the intestine of the pig which is the intermediate host there form round larvæ which possess six hooks These are called oncospheres By means of the hooks, the oncosphere penetrates into the wall of the intestine and is transformed into a later stage the so called fin The fins gain entrance into the small intestine of man by the ingestion of insufficiently cooked pork and there develop into the band worm or Taenia solium In rare cases due to antiperistaltic movements associated with vomiting self infection because of lack of cleanliness or infection from external sources, man becomes an intermediate host for the band worm The oncosphere when present in the human organism develops into a fin and lodges in various organs At this stage, it is termed the cysticercus The organisms are found in the brain eye muscles heart abdominal cavity spinal cord, and breast

The number of cysticercus inclusions in the human organisms varies widely from a few to many hundreds. In one instance over 600 calcified cysticercus inclusions were clearly visualized on the roentgenogram. Each inclusion is in the form of a vesicle filled with fluid. After the death of the parasite it becomes spindle shaped or oval. The longitudinal axis of the worm lies in the direction of the muscle fibers. In the muscle the inclusions tend to be oblong, while in the breast and other soft structures they are rounded. The usual size is 2 to 9 mm. in length and 0.5 to 2 mm. wide. It is believed that the worm lives for about five or six years in the human body. After its death it is resorbed or organized and subsequently becomes calcified.

The calcified bodies have been described in the muscles and the subcutaneous tissues. In no case have they been seen in the central nervous system, the brain, the eye, or the visceral organs. Cysticercus inclusions which have become calcified present no danger to the patient. There is no basis to assume that the parasites localized in the muscles have undergone calcification while others have remained alive. In cases with multiple inclusions the diagnosis is not difficult. When solitary, they are easily confused with phleboliths or similar densities. Trichinosis can be excluded as the trichinae measure less than 1 mm. in diameter and are too small to be demonstrable by roentgen methods. In cases with multiple inclusions the muscles appear to have been sprinkled with sand. Biopsy is essential for accurate diagnosis. Although the condition is not a source of danger to the patient, it is important that the diagnosis be established in order to avoid confusion with other calcific densities which may be of more serious import.

Calcifications of the Spleen

Calcifications of the spleen may involve the capsule or the parenchyma. Those of the capsule may be due to trauma or inflammation, the so-called perisplenitis. The calcific depositions may assume large proportions and cover a large portion of the spleen. In calcifications of the parenchyma two are particularly noteworthy: phleboliths and those due to tuberculosis. Phleboliths are small, usually measuring a few millimeters to about a centimeter in diameter. They are sharply defined and occur singly or in groups. In some instances there are concentric onion peel rings or layers with a zone of increased radiance in the central portion. Tuberculous calcifications tend to be minute and have polyhedral outlines. They are solitary or multiple and may present increased density in the central portion due to calcified central caseation. There is usually evidence of tuberculosis elsewhere in the body. Diagnosis cannot be established from the roentgen manifestations alone, the history and clinical findings being essential. Calcified cysts due to echinococcus may attain a size of several centimeters. Calcification may ensue after local necrosis due to abscesses or infarcts. Syphilitic gumma, cavernous angioma and parasites may cause calcifications or incrustations in the vessels of the spleen. Aneurysm of the splenic artery may be manifested by the presence of a ring-like or mottled area of calcific density in the region of the spleen.

BONE CHANGES PRODUCED BY WOOD IMBEDDED IN THE SOFT TISSUES

In 1952 Maylahn under the title "Thorn induced Tumors of Bone" recorded 3 cases in which thorns of plants imbedded in the soft tissues produced bone changes very similar to those in osseous tumors. He states that, while there are numerous references in the literature describing the reactions of bone to metal, a review of the literature for the past thirty years *yields no reports on the reaction of bone to organic foreign material*. The introduction of catgut into or near a bone while relatively frequent, does not result in a bone reaction. Maylahn states that to his knowledge, only thorns can produce a gross change in bone and suggests that experimental investigation would have to be performed to ascertain whether this type of osseous change is unique to thorns. In two cases recently observed at the Boston City Hospital, changes similar to those described by Maylahn were produced by the presence of a fragment of wood in the soft tissues. After removal of the imbedded foreign body, the bone changes regressed and there was a return to a normal appearance. The first case was in a girl age nine years. She entered the accident ward because of an injury to the forearm sustained in a fall. A laceration on

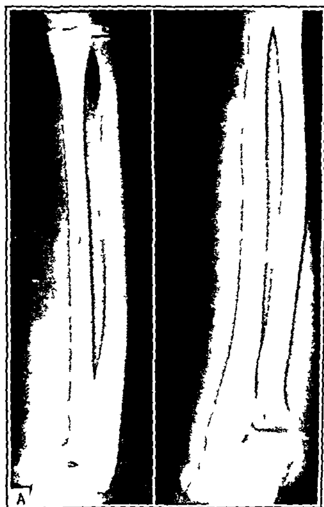


FIG 391 —(*Legend and Illustration continued on opposite page*.)

the forearm was noted X ray examination revealed no bone changes and there was no evidence of a foreign body A splint was applied A week later she returned with swelling and tenderness of the lower part of the forearm anteriorly Penicillin was administered A month later the swelling was still present and there was considerable tenderness The wound was discharging pus Seven weeks after the injury roentgen study showed periosteal reaction The diagnosis of foreign body reaction was made At operation a fragment of wood measuring 5 cm × 2 cm was found deeply imbedded in the muscle adjacent to the ulna and was easily removed Following the operation the swelling and tenderness subsided and the wound healed Four months after the operation the patient was again examined The periosteal reaction had resolved

The second case was that of a boy eleven years old The patient fell from a tree a branch piercing the upper anterior part of the right forearm X ray examination revealed no bone injury or opaque foreign body In the accident ward several small fragments of wood were removed from the wound Eleven weeks later the patient returned because of pain and swelling in the upper part of the forearm and limitation of motion of the elbow X ray examination revealed new bone formation adjacent to the radius in its upper third (Fig 398) The diagnosis of foreign body reaction was made At operation a fragment of wood about 2 cm × 1 cm was found adjacent to the bone Within a few days after removal of the foreign

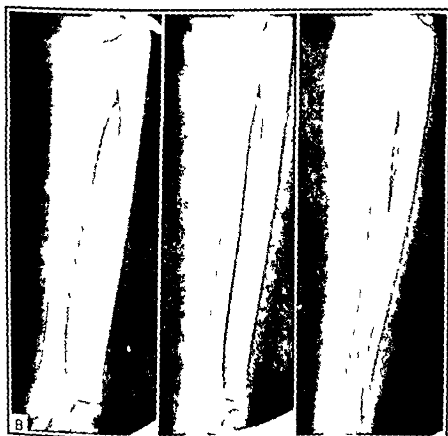


Fig 397 Bone Changes Produced by Wood in the Soft Tissues A Roentgen study seven weeks after injury to the forearm There is extensive periosteal reaction with new bone formation along the lower third of the diaphysis of the ulna A fragment of wood measuring 5 cm × 2 cm imbedded in the soft tissues is not demonstrable roentgenographically B Roentgenogram four mo[illegible]fter removal of a fragment of wood from the forearm The area of calcific density in the [illegible] has disappeared and the periosteal reaction has resolved almost completely

body the soft tissue inflammation had resolved The bone changes subsided and re examination three months later showed no abnormalities in the bones The condition is unusual and the roentgen manifestations are not characteristic A careful history is essential since the foreign body is not visible on roentgen examination The treatment is dependent on correct diagnosis and removal of the foreign body, which results in a complete cure



Fig 398 Bone Changes Produced by Wood Imbedded in the Soft Tissues *A* Oblique view *B* Lateral projection Eleven weeks after the injury There is extensive new bone formation adjacent to the anterolateral aspect of the upper third of the diaphysis of the radius The imbedded fragment of wood is not visualized

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समुद्रस्य गम्भीरं नैव शक्यं चिकित्सितम् ।
वक्तुं निरवशेषेण श्लोकानामयुतैरपि ॥

—सुश्रुत संहिता

'The Science of Medicine is fathomless like the sea and can not be exhaustively narrated in thousands of couplets'

—SUSHRUT SAMHITA